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TUMORS OF THE RETROPERITONEUM MESENTERY AND PERITONEUM

Lauren V. Ackerman, M. D.

ARMED FORCES INSTITUTE OF PATHOLOGY

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ATLAS OF TUMOR PATHOLOGY

Section VI—Fascicles 23 and 24

TUMORS OF THE RETROPERITONEUM MESENTERY AND PERITONEUM

by

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TUMORS OF THE RETROPERITONEUM, MESENTERY, AND PERITONEUM

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Fig. 1

LIPOSARCOMA

(Figures 1 and 2 are from the same case)

Figure 1. Retrograde pyelogram. Extreme displacement of stomach and ureter in a patient with a large retroperitoneal liposarcoma. A. F. I. P. Acc. No. 219023–33.

Figure 2. Lateral roentgenogram of the patient whose pyelogram is shown in figure 1. A. F. I. P. Acc. No. 219023–34.

Fig. 2

TUMORS OF THE RETROPERITONEUM, MESENTERY, AND PERITONEUM

TUMORS OF THE RETROPERITONEUM

(Originally planned as Fascicle 24)

INTRODUCTION

Definition

The retroperitoneal space is that indefinite area in the lumbar and iliac region which lies between the peritoneum and the posterior parietal wall of the abdominal cavity. It extends upward to the twelfth rib and twelfth thoracic vertebra and downward to the base of the sacrum and the iliac crest. The lateral margins correspond to the lateral borders of the quadratus lumborum muscles. The floor of the space is formed by the quadratus lumborum and psoas major muscles, and extends from the lateral lumbocostal arch above to the iliolumbar ligament below. The space contains loose areolar fatty tissue, through which pass the ureters, nerves and ganglia, the renal vessels, and the spermatic vessels in the male and the ovarian vessels in the female. The retroperitoneal tissue abuts against the inferior vena cava on the right and the aorta on the left. There are numerous lymph nodes in this region: iliac lymph nodes in the iliac fossa and many lymph nodes along the aorta. The peritoneum covering the apposed surfaces of the rectum and bladder forms the rectovesical cul-de-sac (pouch of Douglas), which extends to the uppermost margin of the rectovesical fascia bridging the extraperitoneal portions of the rectum and bladder.

Scope

This fascicle includes tumors arising primarily from tissues in this retroperitoneal area; it does not include any discussion of primary neoplasms arising from organs that are partially or completely retroperitoneal such as the pancreas, kidneys, and adrenals, or of tumors primarily or secondarily involving bone and spinal cord. The fascicle, therefore, will not include ependymomas (Hundling), giant cell tumors, chondrosarcoma, and other neoplasms of this region that may invade the retroperitoneal zone. Chordomas of the sacrococcygeal region are included. The discussion briefly mentions metastatic tumors.

Primary retroperitoneal malignant neoplasms represent a variegated rare group. There is a vast literature on retroperitoneal tumors. Stout has collected the largest series of benign and malignant retroperitoneal tumors, based primarily on surgical material. The incidence of various types of tumors in his

series (tables I and II) is not necessarily representative of the true incidence, for a large proportion of these tumors were referred to Stout for diagnosis. It is unfortunate that in the literature gross and microscopic descriptions are often inadequate for diagnosis. There are many single cases reported, and usually there is no follow-up. At times the photomicrographs demonstrate an eloquent incorrect diagnosis. The rarity of these retroperitoneal neoplasms precludes any breakdown into relative frequency, and consistent lack of follow-up and too often incorrect diagnoses render it impossible for any predictions to be made concerning prognosis. This fascicle therefore represents merely an introduction to this complex area and, we hope, a stimulation to more careful classification of neoplasms arising there, as well as to more thorough follow-ups.

Table I

BENIGN RETROPERITONEAL TUMORS IN LABORATORY OF SURGICAL PATHOLOGY, COLUMBIA UNIVERSITY, 1905 to 1951 INCLUSIVE*

	Retroperitoneal	Mesenteric	Omental
Adenoma	1	.....	.....
Papillary cystadenoma	2	.....	.....
Lymphangioma	.....	3	.....
Lipoma	4	2	5
Xanthogranuloma	5	.....	.....
Neurilemoma	1	.....	.....
Neurofibroma	6	1	.....
Ganglioneuroma	4	.....	.....
Granular cell myoblastoma	11	.....	.....
Dermoid cyst	2	.....	.....
Leiomyoma	6	1	4
Paraganglioma (inactive)	2	.....	.....
Pheochromocytoma (active)	14	.....	.....
Cyst of caudal gut	17	.....	.....
Total	75	7	9

*Courtesy Arthur Purdy Stout, M. D.

General Characteristics

There are certain general characteristics common to many of the retroperitoneal tumors. Because the retroperitoneal space is rather extensive, and adjacent organs are easily displaced rather than invaded, many of the neoplasms grow to a large size before giving any clinical manifestations. Fre-

Table II

MALIGNANT RETROPERITONEAL TUMORS IN LABORATORY OF SURGICAL PATHOLOGY, COLUMBIA UNIVERSITY, 1905 to 1951 INCLUSIVE*

	Retroperitoneal	Mesenteric	Omental
Sympathicoblastoma	18	.....	.....
Malignant schwannoma	3	.....	.....
Lymphosarcoma	51	2	2
Hodgkin's disease	26	.....	.....
Plasmocytoma	.....	1	.....
Leukemia	2	.....	.....
Fibrosarcoma	4	6	.....
Liposarcoma	35	2	**2
Leiomyosarcoma	29	2	4
Hemangiopericytoma	18	1	2
Rhabdomyosarcoma	5	.....	.....
Mesenchymoma	5	.....	.....
Myxoma	3	.....	.....
Teratoma	6	.....	.....
Embryonal carcinoma	24	.....	.....
Suprarenal cortical carcinoma	1	.....	.....
Metastatic carcinoma of unknown origin	13	.....	.....
Malignant melanoma (metastatic)	1	.....	.....
Chordoma	2	.....	.....
Dysgerminoma	1	.....	.....
Undiagnosed tumor	18	1	4
Total	265	15	14

*Courtesy Arthur Purdy Stout, M. D.
**3 more attached to kidney capsule.

quently, when the patient is first seen, there is an indefinite abdominal mass the exact nature of which is obscure, and the diagnosis is not made until the time of surgical investigation. The operation is often complicated by the large size of the tumor and by its attachment to vital structures; therefore, operative mortality is high and complete removal difficult. It has been demonstrated in numerous instances, particularly by urologists (Sweetser), that certain radiographic findings may be helpful in demonstrating a tumor to be retroperitoneal (Windholz): these large masses displace the ureters, the bowel, and the stomach (figs. 1, 2). These pressure phenomena may result in various paresthesias, with

radiation of pain often to the thighs and legs, and with blockage and thrombosis of veins such as the iliacs or inferior vena cava, which in turn result in edema of the lower extremities and occasionally ascites. These tumors may extend into the mesentery and the large and small intestines, as well as over the promontory of the sacrum, into the pelvic connective tissues, and into the sigmoid mesocolon. They may grow through the inguinal canal into the labium and buttock (Delamater) and even through the obturator foramen. They frequently obstruct ureters and can cause hydronephrosis and renal failure.

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Hundling, H. W. Ventral tumors of the sacrum. Surg., Gynec. & Obst., 38:518–533, 1924.

Stout, A. P. Personal communication.

Sweetser, T. H. Retroperitoneal tumors influencing the kidneys and ureters. J. Urol., 47:619–631, 1942.

Windholz, F. Roentgen diagnosis of retroperitoneal lipoma. Am. J. Roentgenol., 56:594–600, 1946.

Classification

There has never been a satisfactory classification of the retroperitoneal tumors. The following is suggested as a tentative classification:

I. Tumors of mesodermal origin
  A. Tumors arising from adipose tissue
    1. Benign—lipoma
    2. Malignant—liposarcoma
  B. Tumors arising from smooth muscle
    1. Benign—leiomyoma
    2. Malignant—leiomyosarcoma
  C. Tumors arising from connective tissue
    1. Benign—fibroma (not reported)
    2. Malignant—fibrosarcoma
  D. Tumors arising from striated muscle
    1. Benign—rhabdomyoma (not reported)
    2. Malignant—rhabdomyosarcoma
  E. Tumors arising from lymph vessels
    1. Benign—lymphangioma
    2. Malignant—lymphangiosarcoma (unknown)
  F. Tumors arising from blood vessels
    1. Hemangioma and angiosarcoma
    2. Hemangiopericytoma, benign and malignant
  G. Tumors arising from primitive mesenchyme
    1. Myxoma—myxosarcoma
  H. Tumors of uncertain origin
    1. Xanthogranuloma

II. Tumors of neurogenous origin
  A. Tumors of nerve sheath origin
    1. Nonencapsulated neurofibroma
    2. Encapsulated neurilemoma
    3. Malignant schwannoma
  B. Tumors arising from sympathetic nervous system
    1. Ganglioneuroma
    2. Sympathicoblastoma
    3. Neuroblastoma
  C. Tumors arising from heterotopic cortical adrenal and from chromaffin tissue
    1. Carcinoma arising from cortical adrenal tissue
    2. Malignant nonchromaffin paraganglioma
    3. Paraganglioma
    4. Functioning pheochromocytoma
III. Remnants of renal blastema
IV. Tumors arising from embryonic remnants
  A. Benign and malignant teratomas
  B. Chordomas
V. Metastatic malignant tumors

As can be seen from the classification, there are many different types of tumors which are naturally related to the great variation in the types of tissues found in the retroperitoneal space. Complete descriptions of practically all of these tumors may be found in individual fascicles: for instance, Stout has discussed the soft tissue and neurogenous sarcomas,* Karsner the adrenal neoplasms,** and Willis the teratoma.***

We have tried to avoid repetition of material presented by other authors, but the various tumors of this area are illustrated here, and their important gross and microscopic features briefly described; any difference in appearance because of location has been stressed.

TUMORS OF MESODERMAL ORIGIN

TUMORS ARISING FROM ADIPOSE TISSUE

Benign fatty tumors, **lipomas**, are infrequent. DeWeerd and Dockerty reported 43 retroperitoneal fatty tumors of which 28 were malignant and 15

*Fascicle 5, "Tumors of the Soft Tissues," and Fascicle 6, "Tumors of the Peripheral Nervous System."

**Fascicle 29, "Tumors of the Adrenal."

***Fascicle 9, "Teratomas."

PLATE I

LIPOSARCOMA

A. Enormous retroperitoneal bosselated liposarcoma, in situ. This case illustrates the difficulty of adequately resecting such a neoplasm. A. F. I. P. Acc. No. 219023–26.

B. Lobulated bright yellow liposarcoma. A. F. I. P. Acc. No. 219023–27.

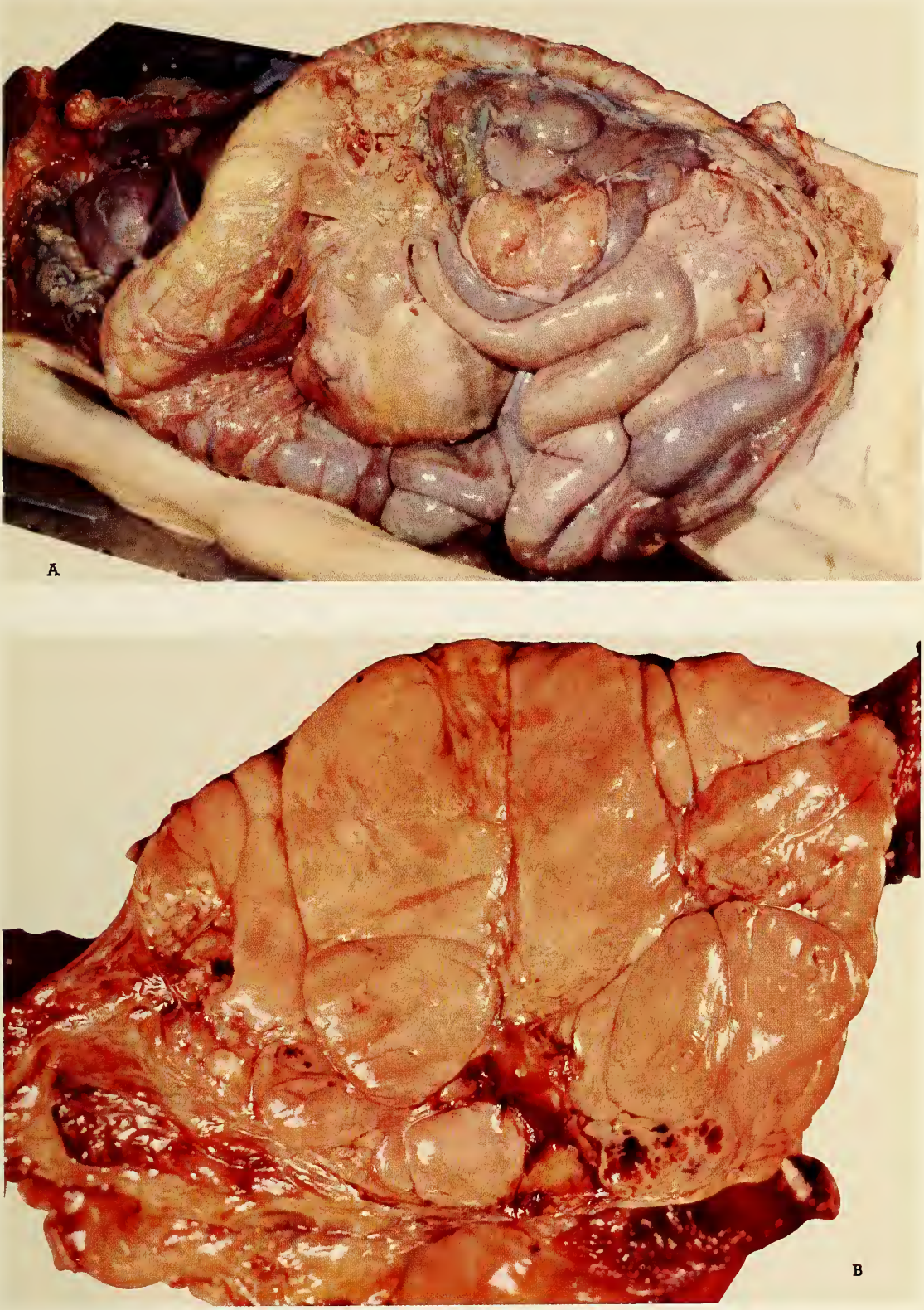

PLATE I

were benign. Of 21 of the malignant group that had been surgically resected, only one was apparently cured.

Liposarcomas are probably the most common neoplasms of the retroperitoneal region. These tumors grow to an extremely large size: the record weight was reported by Delamater with a lipoma of about 275 pounds; Wells reported one of 69 pounds. These tumors occur about equally in males and females, usually in the older age groups, but they can appear in children (Kretschmer). These liposarcomas form lobulated masses of variable consistency (pl. I–A, B). Their surface may have the appearance of cerebral convolutions and on section may resemble brain tissue. More than half have a mucoid yellow slimy appearance so that grossly they are mistaken for myxoma. Such large neoplasms may form multiple masses; and, although they are frequently easy to shell out, they often have fine ramifications that are impossible to remove (figs. 3, 4). These fatty malignant tumors have a predilection for the perirenal area. Their intimate relation to the kidney may necessitate its removal. At times, liposarcomas of the retroperitoneal area are associated with other independent liposarcomas (Ackerman; Siegmund) or with benign fatty tumors (Starkloff, Saxton, and Johnson). Microscopically, these tumors show all the variations that can be seen in any group of liposarcomas (fig. 5). Some are well differentiated, others poorly differentiated; and the gradations from well differentiated to poorly differentiated may be found in the same tumor. The well differentiated tumors are frequently incorrectly diagnosed as lipomas, and often the medical literature has had reports of a large lipoma of the retroperitoneal space that was removed but later recurred and became malignant (Hosemann; von Wahlendorf). In practically all instances, however, if the tumor is malignant it is malignant from the start, and only under exceptional circumstances does it arise from a lipoma (Wright). The treatment of these tumors is surgical; but it is often difficult to remove them completely, and therefore recurrences appear. These liposarcomas, however, are to some extent radiosensitive (Stout, 1944); this may be related to their histogenesis (Wells).

TUMORS ARISING FROM SMOOTH MUSCLE

Leiomyomas are extremely rare, unless those of uterine origin are considered; the latter can form large retroperitoneal masses. Lindeman reported a 49-pound cystic leiomyoma arising from a pedicle behind the left uterine horn between the layers of the left broad ligament.

Leiomyosarcomas appear to be next in frequency to tumors of fatty origin. The malignant smooth muscle tumors often form large masses which may infiltrate and become fixed to surrounding structures, and may have pseudoencapsulation and central cystic degeneration (Lumb; figs. 6, 7). They are firmer than the lipsarcoma and may present a surface pattern suggesting smooth muscle

LIPOSARCOMA

(Figures 3 and 4 are from the same case)

Figures 3 and 4. Large retroperitoneal liposarcoma growing in the region of the kidney. Note lobulated external surface and mucoid appearance on gross section. "A" indicates kidney attached to lower pole of specimen. A. F. I. P. Acc. Nos. 219023–35 and 219023–36.

Figure 5. Note cellularity, good differentiation, and compression of the nucleus to a crescentic shape by cytoplasmic fat. × 555. (From the Armed Forces Institute of Pathology.) A. F. I. P. Acc. No. 168137–1.

Fig. 3

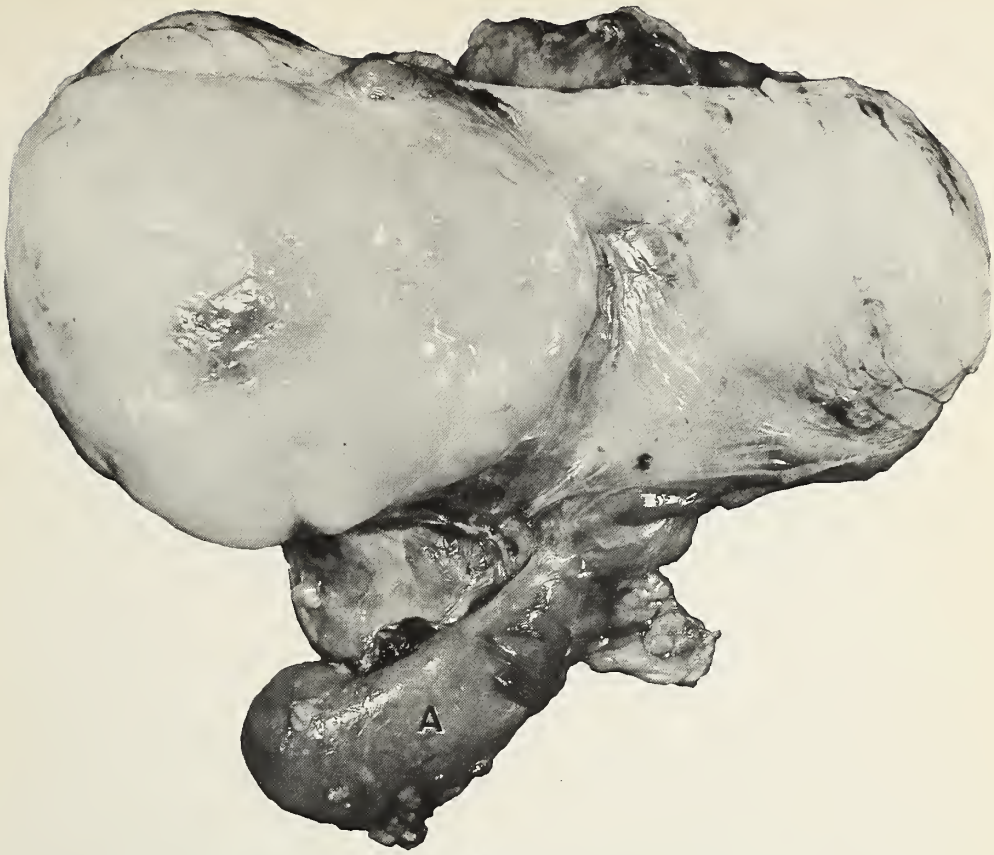

Fig. 4

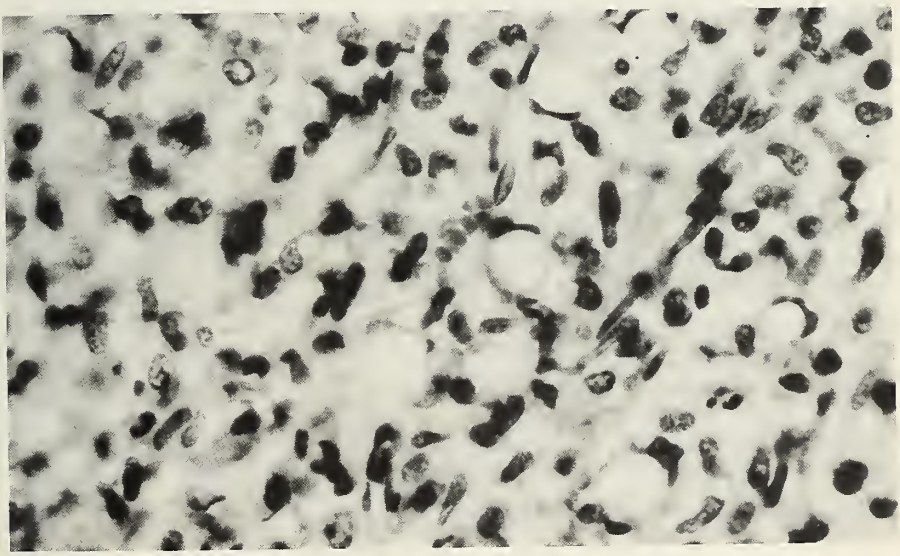

Fig. 5

LEIOMYOSARCOMA

(Figures 8 and 9 are from the same case)

Figure 8. Microscopic section of a leiomyosarcoma. Note the alignment of the nuclei and their rounded ends. × 650. A. F. I. P. Acc. No. 97535–1.

Figure 9. Higher magnification of cells of tumor illustrated in figure 8. × 915. A. F. I. P. Acc. No. 97535–2.

Fig. 8

Fig. 9

RHABDOMYOSARCOMA

(Figures 10–12 are from the same case)

Figure 10.* Gross specimen of rhabdomyosarcoma arising retroperitoneally from the psoas major muscle. Note hemorrhagic appearance and conformation to the shape of the muscle. A. F. I. P. Acc. No. 219023–41.

Figure 11. Note the long cytoplasmic processes. × 600. A. F. I. P. Acc. No. 219023–61.

Figure 12.* Note prominent nuclei and long cytoplasmic processes. No clearly visible cross striations could be identified. × 300. A. F. I. P. Acc. No. 219023–42.

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*From Ackerman, L. V., and del Regato, J. A. Cancer. Diagnosis, Treatment and Prognosis. St. Louis: C. V. Mosby Co., 1947. Figures 717B and C are our figures 10 and 12.

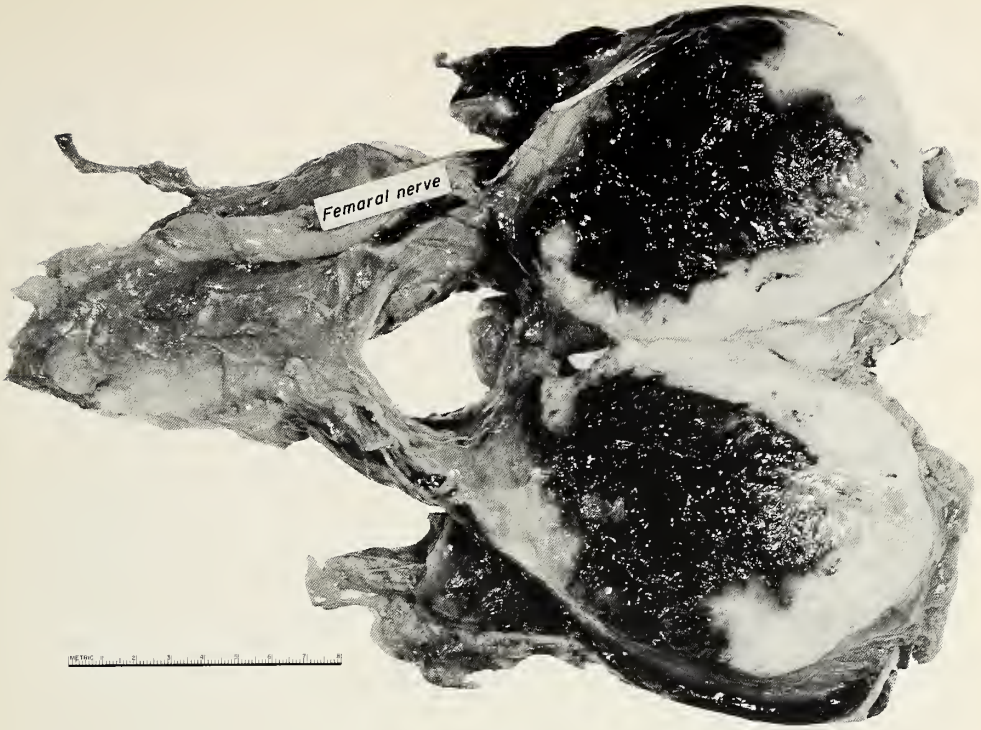


Fig. 10

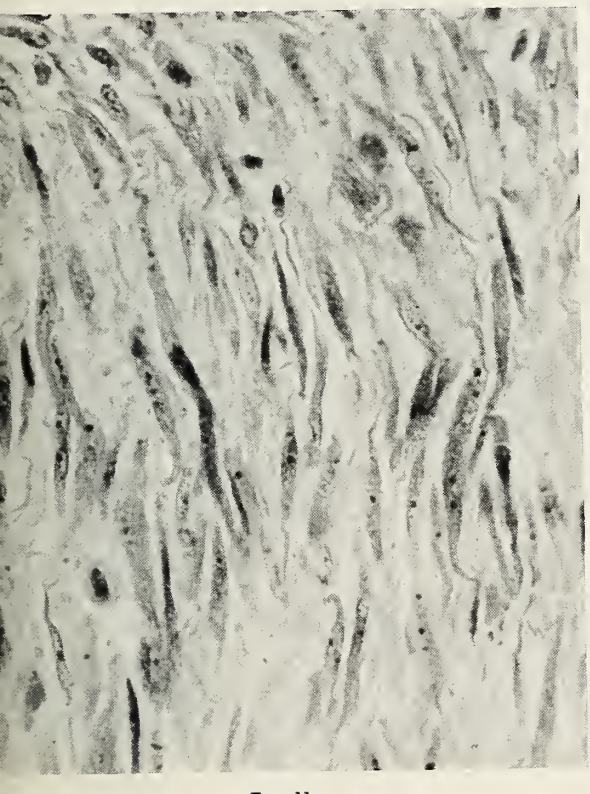


Fig. 11

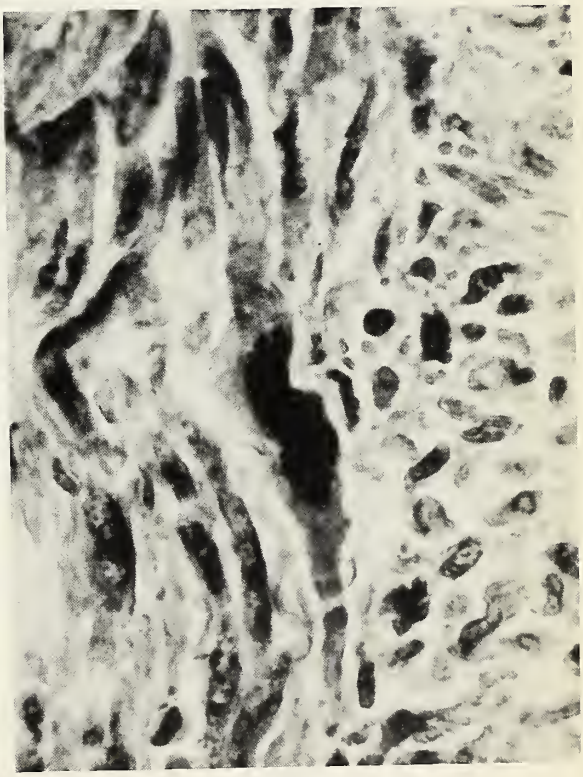


Fig. 12

RETROPERITONEAL FIBROSARCOMA

Figure 13. Large retroperitoneal fibrosarcoma arising between the kidney and the adrenal. Cut section demonstrates a pseudoencapsulated homogeneous surface with central necrosis. (Contributed by Dr. J. P. Thomas, Baylor University School of Medicine, Houston, Texas.) A. F. I. P. Acc. No. 219023–40.

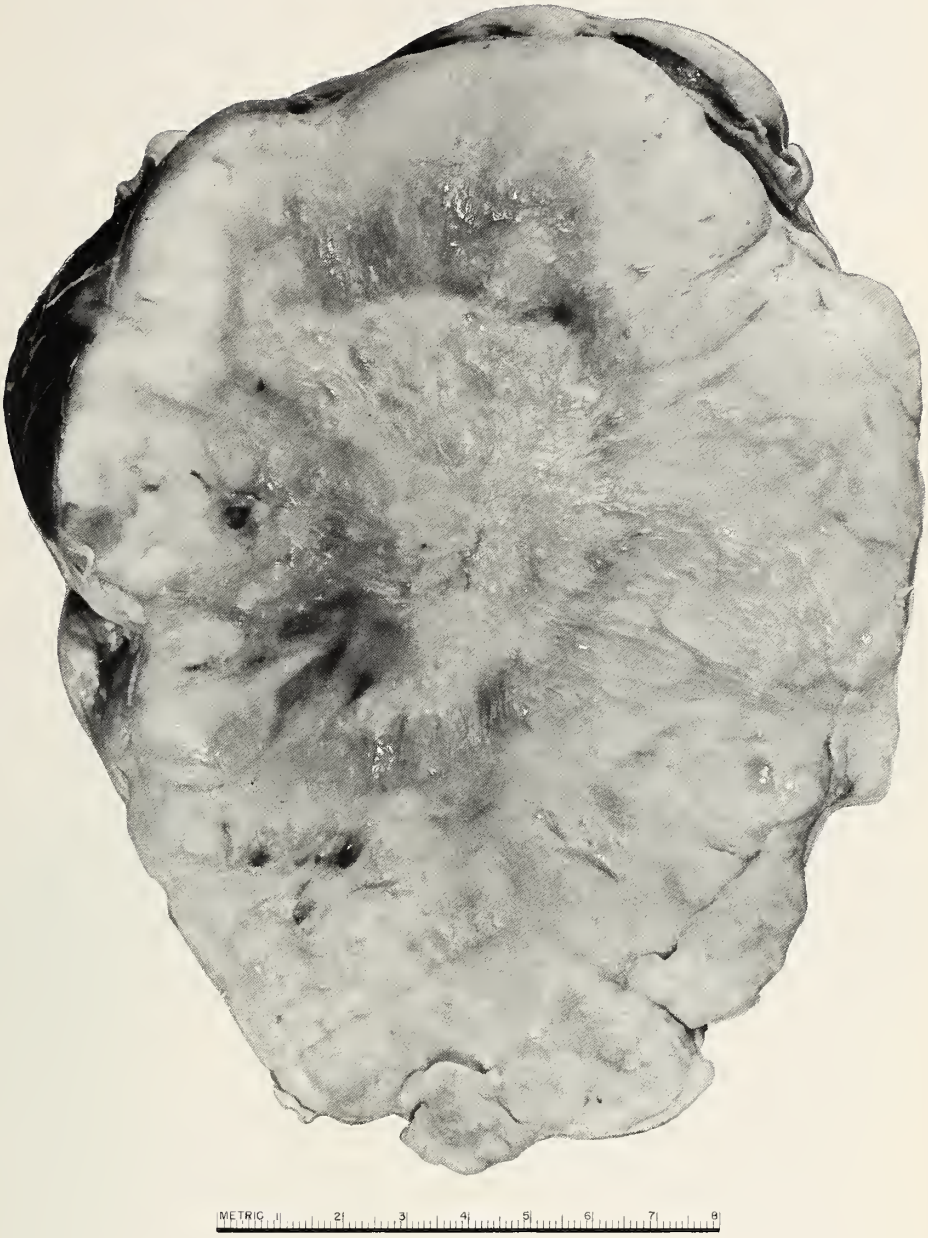


Fig. 13

LYMPHANGIOMA

Figure 14.* Photomicrograph of a multicystic lymphangioma. Malignant change in such a lesion has not been reported. × 15. A. F. I. P. Acc. No. 58611–1.

Figure 15.* Photomicrograph of another multicystic lymphangioma demonstrating lymphoid follicles with dilated lymphatic channels lined by endothelial cells. × 48. A. F. I. P. Acc. No. 91499–1.

*From the Armed Forces Institute of Pathology.

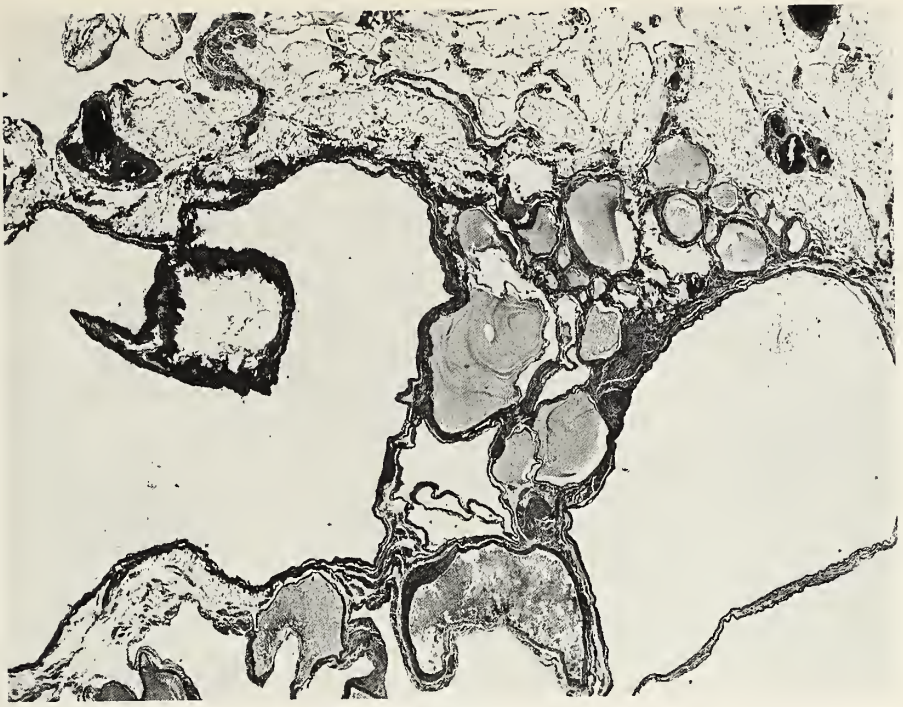


Fig. 14

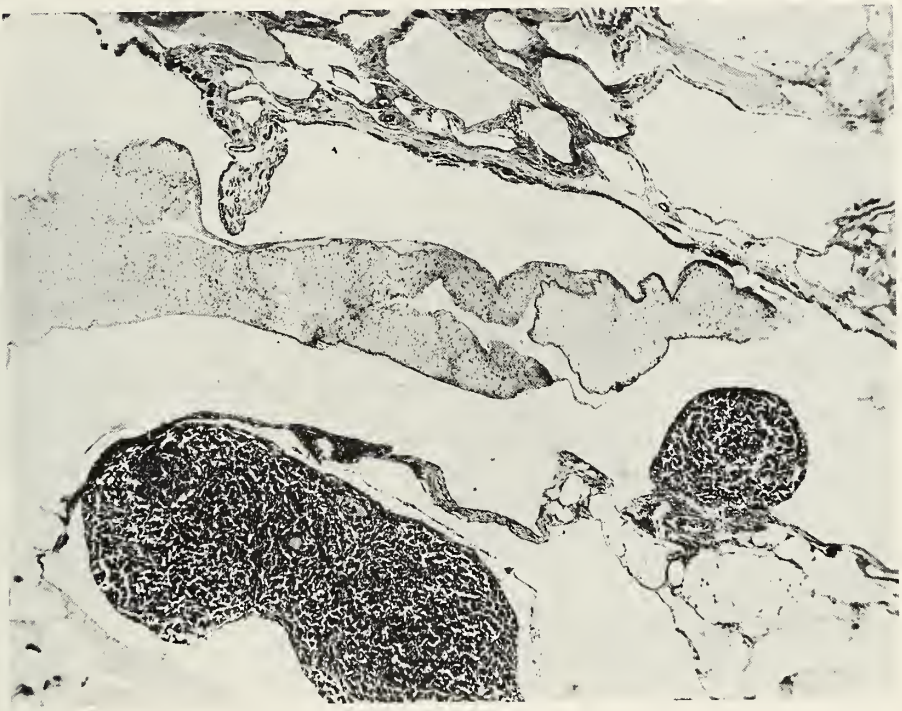


Fig. 15

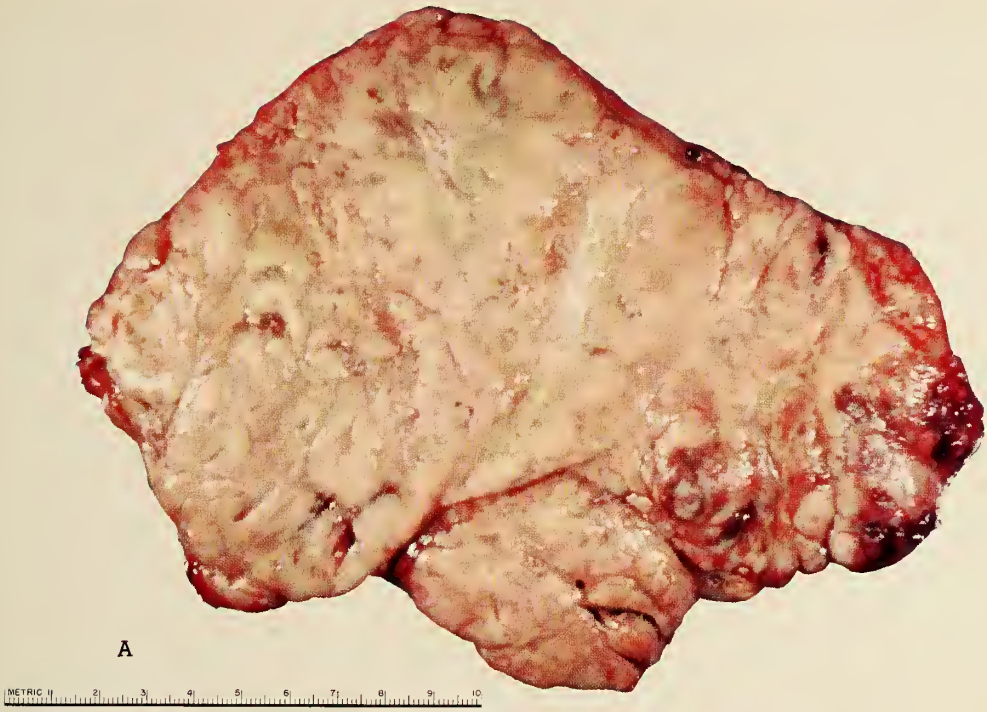
PLATE II

HEMANGIOPERICYTOMA

A. Large homogeneous retroperitoneal hemangiopericytoma. (Contributed by Dr. A. P. Stout, Columbia University, College of Physicians and Surgeons, New York, N. Y.) A. F. I. P. Acc. No. 219023–C2.

ADRENAL CORTICAL CARCINOMA

B. Large retroperitoneal cortical carcinoma of the adrenal. A. F. I. P. Acc. No. 219023–28.



A

B

PLATE II

fluid. There has always been an argument as to whether these cysts are new growths or develop from varicosities of newly formed lymph vessels. These cysts sometimes contain endothelial spaces filled with blood, and often there are a few collections of smooth muscle cells. They can be found in the mesocolon (Kayser) and in the mesentery (Lubitz and Flynn). Gerster's extensive bibliography indicates the important references and summarizes succinctly all case reports. Kayser (1914) found 15 lymphangiomas of the mesocolon which were unilocular cysts.

TUMORS ARISING FROM BLOOD VESSELS

Retroperitoneal **hemangiomas** are rare lesions. Hilse reported a retroperitoneal lipoma with areas of cavernous hemangioma in it. Harris recorded an instance of a large hemangioma which was perirenal and mistaken clinically for a kidney. Millman reported a large one in a man, 32 years of age, lying on the right iliopsoas muscle and displacing the cecum and ascending colon to the midline. It was surgically removed. Ward and Stewart reported a case in a white boy, age five months, with hemangioma of the skin of the buttock and midsacral region. The patient also had a soft tissue mass lying posterior to the coccyx. Attempted surgical removal was unsuccessful. At autopsy there was a large retroperitoneal cavernous hemangioma within the pelvic floor, intimately associated with the bladder (figs. 16, 17). Venous communications between the iliac veins and the tumor were demonstrated.

Angiosarcomas are extremely rare in the retroperitoneal space (Beitzke), and we have not seen a single example. In his review of the literature Stout* found only three references. These tumors are similar to angiosarcoma seen elsewhere.

Stout (1949) has described a vascular tumor designated as a **hemangiopericytoma.** This tumor takes its name from its vascular character and its origin from cells related to Zimmermann's pericytes. Pericytes are cells with contractile ability applied to the outer walls of capillaries and related in some fashion to smooth muscle. In Stout's collection of 25 cases, four were retroperitoneal in origin. Grossly the tumor is often large and vascular and appears encapsulated (pl. II–A). Microscopically it is highly vascular and with conventional stains may exhibit a bewildering pattern (fig. 18). Reticulin stains serve to outline the true nature by bringing out the vascular spaces (fig. 19). The microscopic pattern is not a key to its malignancy. In the retroperitoneal area, complete removal of a hemangiopericytoma is difficult and the prognosis poor.

*Fascicle 5, "Tumors of the Soft Tissues."

CAVERNOUS HEMANGIOMA

(Figures 16 and 17 are from the same case*)

Figure 16. In situ photograph showing anterior surface of bladder. A. F. I. P. Acc. No. 219023–30.

Figure 17. In situ photograph of an extremely large retroperitoneal cavernous hemangioma of the pelvic floor, intimately associated with the bladder and rectum. A: posterior wall of bladder; B: hydroureter (left); C: anterior surface of sigmoid with hemangioma. A. F. I. P. Acc. No. 219023–29.

*From Ward, G. E., and Stewart, E. H., Jr. Retroperitoneal cavernous hemangioma. Am. J. Surg., 80: 470–472, 1950. Figures 2 and 3 are our figures 16 and 17.

Fig. 16

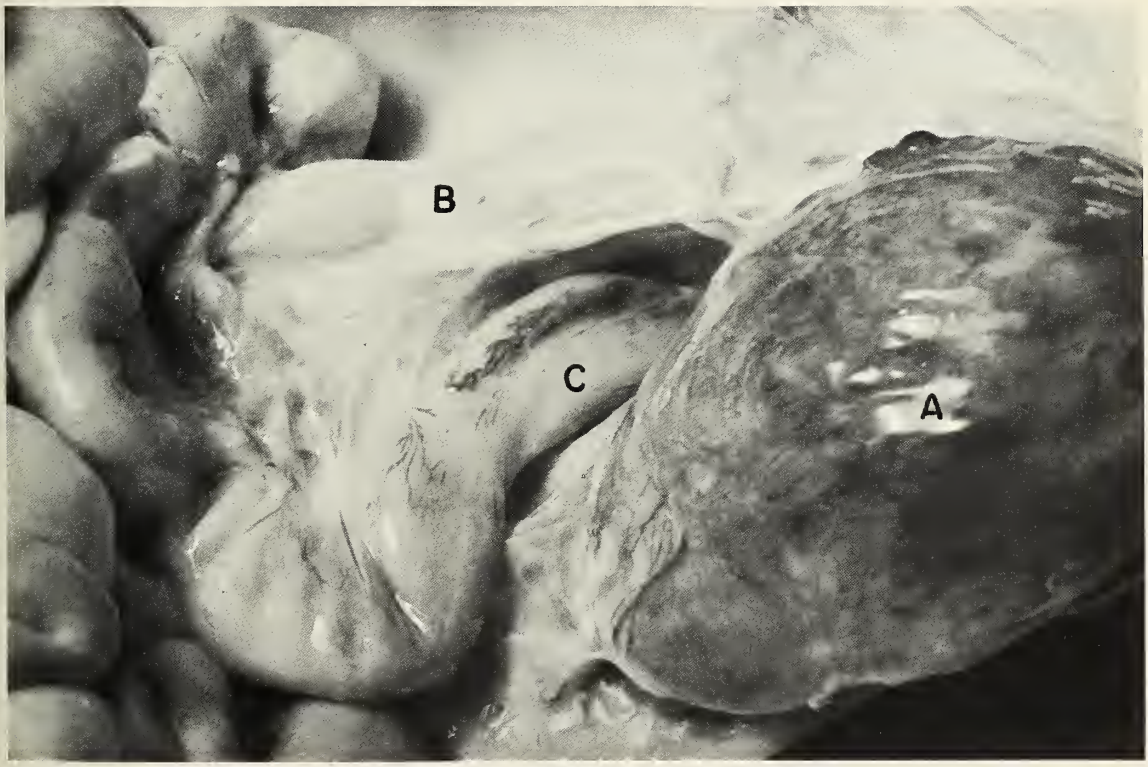


Fig. 17

HEMANGIOPERICYTOMA

(Figures 18 and 19 are from the same case*)

Figure 18. Photomicrograph of typical hemangiopericytoma. Small capillaries are compressed and lined by normal endothelial cells, with proliferation of tumor cells around these capillaries. This relationship cannot be seen on the routine hematoxylin and eosin stain. × 650. A. F. I. P. Acc. No. 219023–43.

Figure 19. This Wilder reticulin stain clearly demonstrates well defined capillary sheaths. Tumor cell proliferation is outside these sheaths. × 650. A. F. I. P. Acc. No. 219023–44.

*Contributed by Dr. Olive Gates, Boston, Mass.

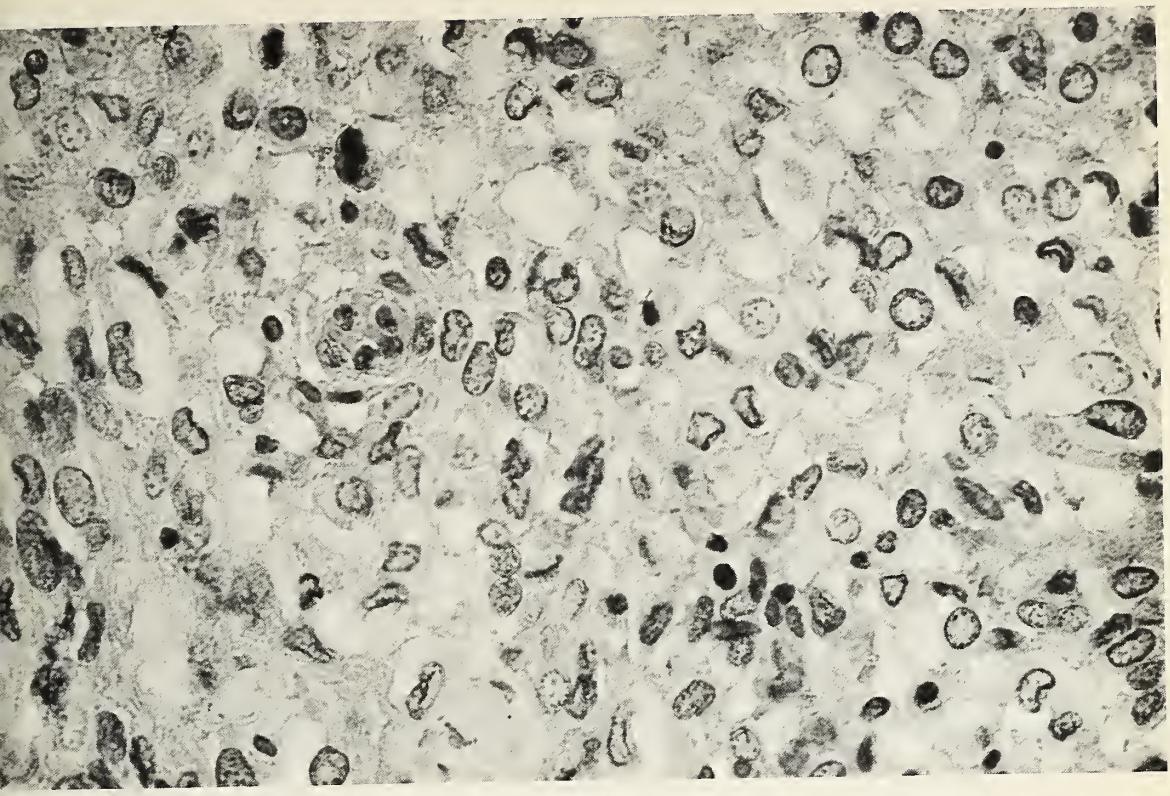


Fig. 18

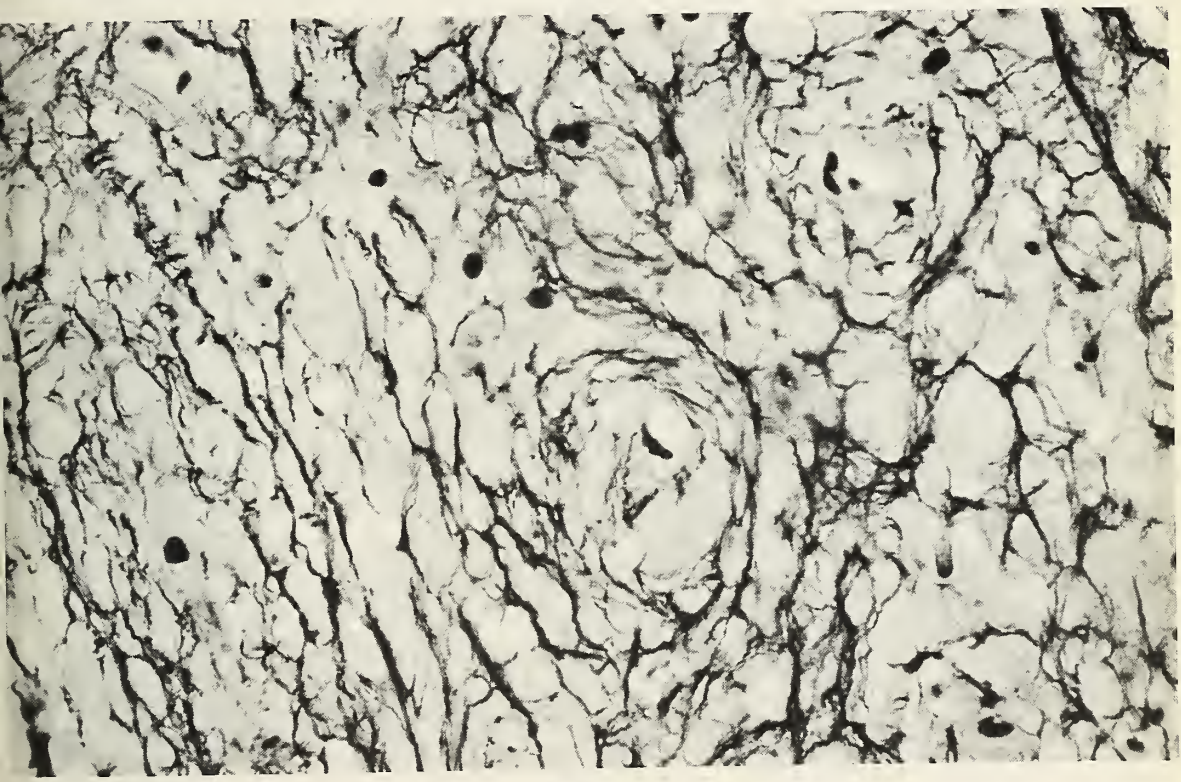


Fig. 19

TUMORS ARISING FROM THE PRIMITIVE MESENCHYME

The **myxoma** (**myxosarcoma**) is rarely found in the retroperitoneal area; it is most common in the heart, skin, subcutaneous and aponeurotic tissues, in certain bones, and in the genitourinary system. In an extensive review of the literature, Stout (1948) found only five retroperitoneal myxomas out of a total of 143. These rare tumors arising from the primitive mesenchyme may grow to extremely large size and infiltrate surrounding tissue, but they never metastasize. It would seem proper to call them malignant on the basis of their tendency to local infiltrative growth and repeated recurrences. Grossly the myxoma may be soft or firm, and on cross section often has a gelatinous quality with areas of cystic change (figs. 20, 21). Microscopically the stellate cells are separated by abundant mucoid material in which hyaluronic acid is found and have fine, tangled reticulin fibers (fig. 22). It is worth while to try to remove these large retroperitoneal tumors, although such a removal often cannot be complete. The time interval between removal and recurrence may be long. In the case of Dixon and Vadheim, a 55-year-old man had a large retroperitoneal myxoma which measured 15 cm. in diameter and weighed 180 gm. The patient was well for eight years. When the tumor recurred it was again resected, and at that time the right colon had to be removed with a small section of the anterior wall of the inferior vena cava.

TUMORS OF UNCERTAIN ORIGIN

Oberling has reported several unusually large tumors which occurred in the retroperitoneal area and which he called **xanthogranuloma.** In the case illustrated, a 31-year-old man had a mass which measured 20 x 8 x 7 cm.; it was quite firm and was removed, only to recur two years later, when it was again removed. There was evidence microscopically of considerable chronic inflammation, prominent fibroblastic proliferation, and collections of cells which contained fat as stained with Scharlach R. There were also sclerotic areas made up of heavy borders of collagen (figs. 23, 24). These fibroblastic areas of proliferation often cause such lesions to be incorrectly diagnosed as fibrosarcoma. The exact pathogenesis and etiology of such lesions are obscure, but it is probable that they are not true neoplasms. There is no evidence that this entity is related to Hand-Schüller-Christian disease. This process is localized and has never been known to metastasize. Treatment should be surgical. In one case seen in a child, surgical removal was inadequate but the tumor regressed further with irradiation.

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MYXOMA

Figures 20 and 21. Gross photograph of a retroperitoneal myxoma in a 2-year-old boy. This tumor lay between the bladder and rectum and measured 7.5 x 4.5 cm. (Case courtesy of Dr. A. P. Stout, Columbia University, College of Physicians and Surgeons, New York, N. Y. Figure 91 from Stout, A. P., Human Cancer, is our figure 21. Philadelphia: Lea & Febiger, 1932.) A. F. I. P. Acc. Nos. 219023–45 and 219023–46.

Figure 22. Photomicrograph of a typical myxoma with stellate cells growing in a loose background. × 900. A. F. I. P. Acc. No. 219023–109.

Fig. 20

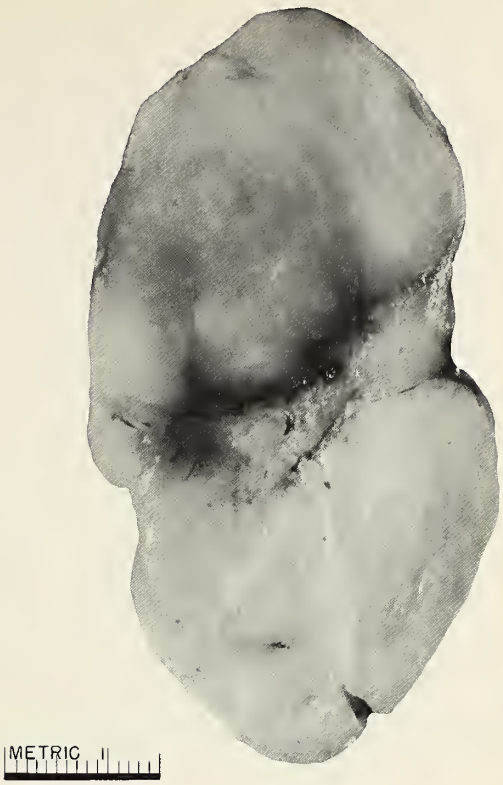

Fig. 21

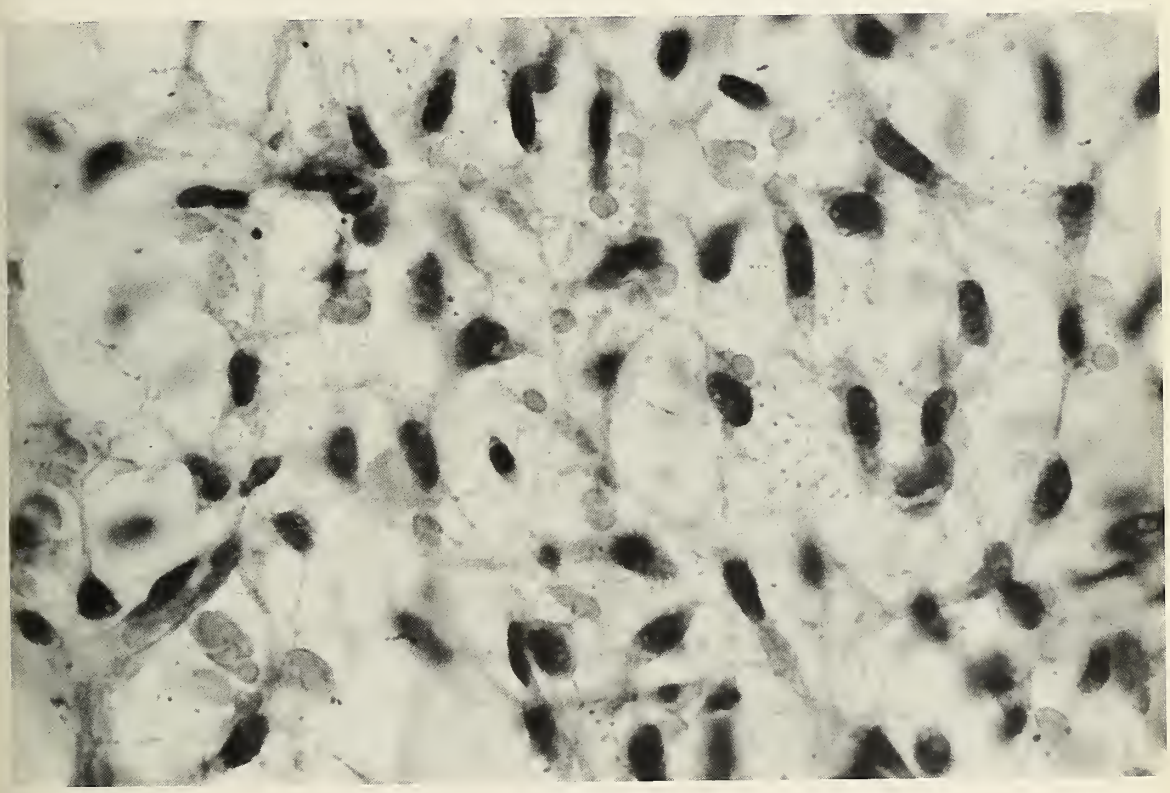

Fig. 22

XANTHOGRANULOMA

(Figures 23 and 24 are from the same case*)

Figure 23. Photomicrograph of a retroperitoneal xanthogranuloma occurring in a 60-year-old man. Varying patterns of connective tissue and large macrophages. × 240. A. F. I. P. Acc. No. 219023–10.

Figure 24. Higher magnification, demonstrating relatively noncellular hyalinized pattern of connective tissue with little cellular infiltration. × 565. A. F. I. P. Acc. No. 219023–12.

*Courtesy of Dr. M. M. Melicow, Department of Urology, Columbia University, College of Physicians and Surgeons, New York, N. Y.

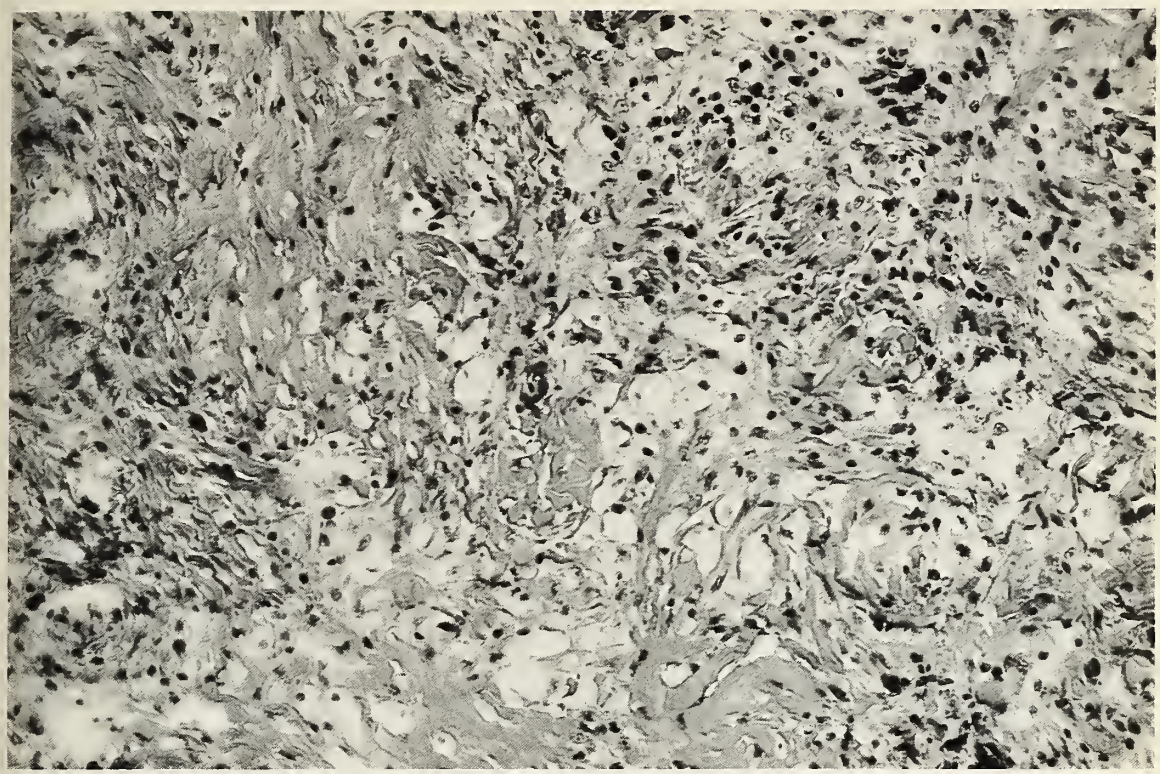

Fig. 23

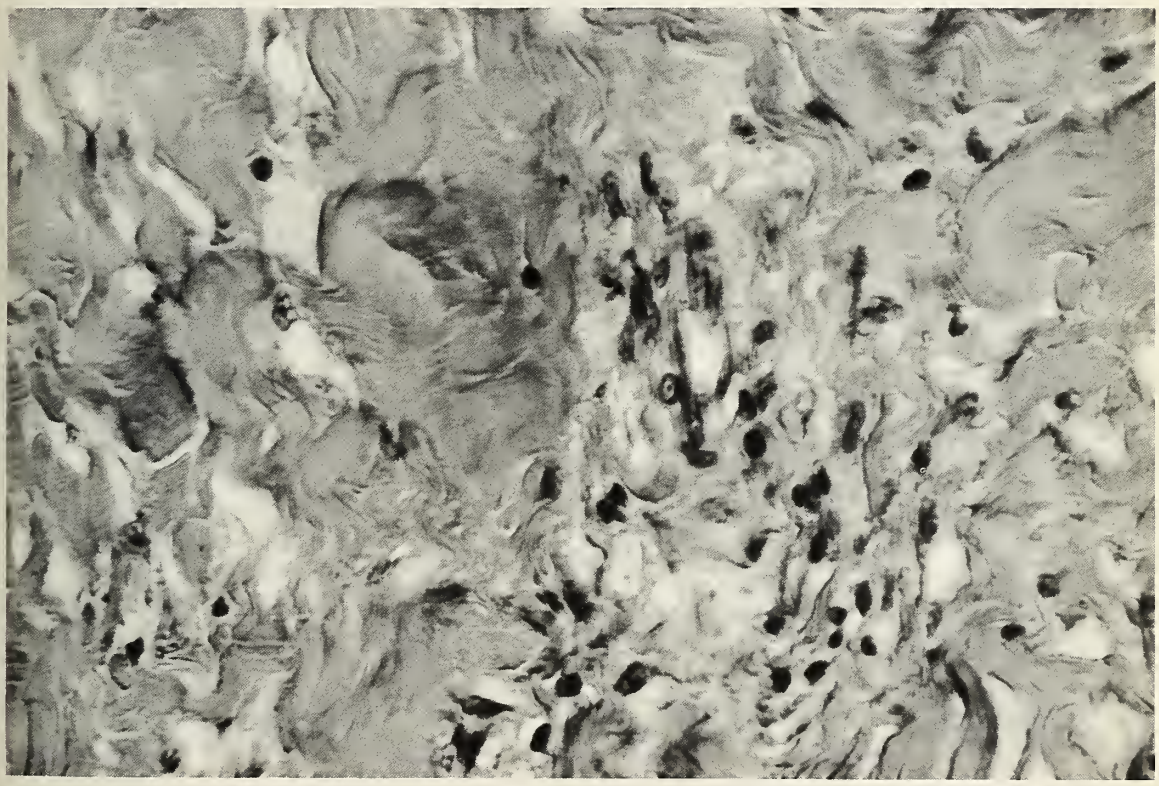

Fig. 24

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TUMORS OF NEUROGENOUS ORIGIN

TUMORS ARISING FROM NERVE SHEATH

Neurofibromas are infrequent in the retroperitoneal area but occasionally are present as a simple independent neoplasm or associated with von Recklinghausen's disease (figs. 25–27).

Neurilemomas are not often found in the retroperitoneal region (Stout, 1935; fig. 28). Neurogenous tumors develop rarely as extradural hourglass neoplasms, growing in the retroperitoneum from within the spinal canal; while other neurogenous tumors develop in the sheaths of retroperitoneal and pelvic nerves (Eichhoff; Korbsh; Pescatori). These tumors may appear in strange places: a cystic tumor described by Erb was attached to the obturator nerve and projected into the obturator canal, and also beneath Poupart's ligament. Another tumor was attached to branches of the sacral plexus, and was mistaken clinically for ectopic gestation (Frank). In another case, the tumor lay behind the rectum pressing against it (Krekeler; also described by Krumbein). In Moreau and Van Bogaert's case the tumor was attached to the crural nerve in the pelvis; and one described by Panà lay behind the bladder and prostate. These tumors may be large, encapsulated, cystic, and may even contain calcium and bone (figs. 28, 29).

We have seen a retroperitoneal neurilemoma attached to the sciatic nerve of a 27-year-old woman. The tumor measured 6 x 8 cm., and at operation was easily separated from the sciatic nerve. It was solid and on section was grayish yellow, with small areas of necrosis. Microscopically the tumor was made up of widely spaced, sinuously curved, and folded reticulin fibers, accompanied at times by elongated thin cells with deep-staining nuclei unmistakably schwannian in nature.

The malignant variant of the neurilemoma, probably best designated as the **malignant schwannoma**, is rare in the retroperitoneal space. Such deep-lying malignant tumors may be accompanied by manifestations of von Recklinghausen's disease. In our only case, the patient, a woman, had a huge tumor with areas of calcification well demonstrated by radiographic examina-

tion (figs. 31, 32). This huge firm tumor weighing 1250 gm. could not be completely removed at operation because it involved the lumbar and sacral plexus. Two months later it was necessary to operate again because the tumor had widely invaded the extradural space. Distant metastases developed in the left humerus, ilium, and pubic bones. Microscopically the evidence of malignant change in a tumor of this nature may be difficult to recognize. This change may be recognized by the increased cellularity; mitotic figures are scarce. It is hard to understand why a tumor with this microscopic pattern can develop wide dissemination (figs. 33, 34).

TUMORS ARISING FROM SYMPATHETIC NERVOUS SYSTEM

Tumors of the sympathetic nervous system arise from the sympathetic ganglia of the retroperitoneal area. **Ganglioneuromas** may also arise in the mediastinum or from the adrenal; they have been well described by Schlumberger,* by Karsner,** and by Stout.*** Within the retroperitoneal area, these tumors (figs. 35, 36), like the other tumors of this region, may grow to a large size. There is some correlation between age and differentiation; generally speaking, undifferentiated tumors occur in children, and the more differentiated tumors in young adults. Stout (1947) collected about 250 cases of ganglioneuromas of the sympathetic nervous system, of which 44 were in the lumbar area, 32 in the pelvic zone, 31 from the suprarenal, and 5 from the mesentery. The tumor reported by Zagarese weighed 6.25 kg. and measured 54 x 42 cm..

In the undifferentiated group, **ganglioneuroblastomas** and **neuroblastomas** are less frequently described. The ganglioneuroblastomas are tumors of the sympathetic nervous system which lie midway between the well differentiated and benign ganglioneuromas, and the poorly differentiated neuroblastomas (figs. 37–40). These ganglioneuroblastomas have a variable microscopic pattern and may show differentiated and undifferentiated elements of the sympathetic nervous system. Of 17 tumors reported by Blacklock, 15 originated in the suprarenal medulla, one in the left superior cervical ganglion, and one in the celiac plexus. The last tumor occurred in a male child, 1 year and 8 months old, and could be called ganglioneuroblastoma. The first evidence of this tumor was a metastatic lesion in the right temporal bone. We have seen a ganglioneuroblastoma in a 6-month-old girl treated by surgery and postoperative irradiation with apparent success, since the baby is still alive 33 months later and without evidence of disease (figs. 38–40). Wittenborg points out that after surgery with postoperative irradiation, cure is often possible. Even

*Fascicle 18, "Tumors of the Mediastinum."

**Fascicle 29, "Tumors of the Adrenal."

***Fascicle 6, "Tumors of the Peripheral Nervous System."

NEUROFIBROMA

(Figures 25 and 26 are from the same case*)

Figure 25. Retroperitoneal encapsulated neurofibroma. A. F. I. P. Acc. No. 271674–1.

Figure 26. Cut surface of tumor illustrated in figure 25. This shows the characteristic grayish white appearance. A. F. I. P. Acc. No. 271674–2.

NEUROFIBROMA

Figure 27. Tumor measured 9 x 7 x 6 cm. It was located in the presacral area of a 24-year-old woman and was complicated by pregnancy. (Figure 2 from Dockerty, M. B. Pregnancy complicated by presacral neurofibroma, one of the so-called Middeldorpf tumors: report of case. Proc. Staff Meet., Mayo Clin., 23: 239–244, 1948. A. F. I. P. Acc. No. 219023–50.

*From the Armed Forces Institute of Pathology.

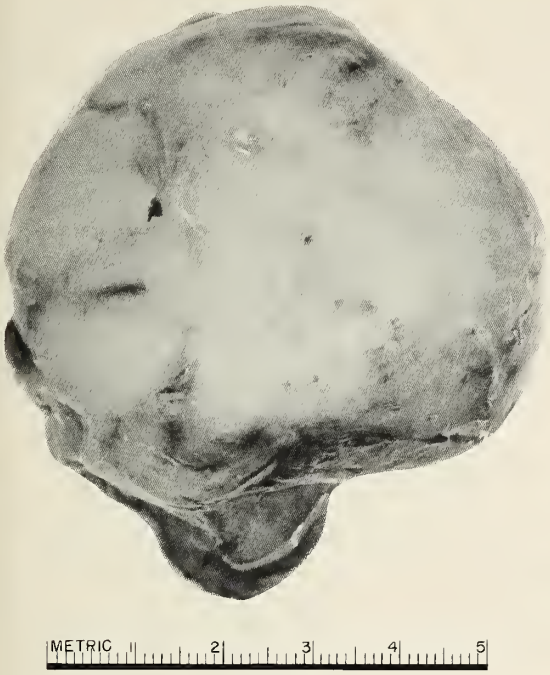

Fig. 25

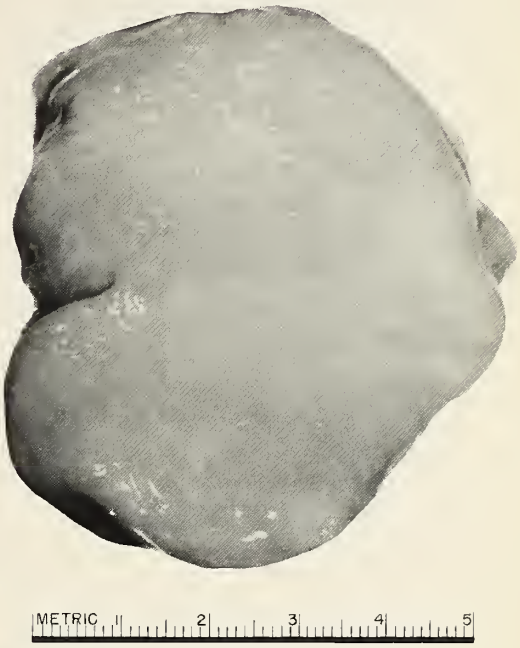

Fig. 26

Fig. 27

NEURILEMOMA

(Figures 28–30 are from the same case*)

Figures 28 and 29. External and cut surfaces of a large retroperitoneal neurilemoma occurring in a 45-year-old woman. Symptoms had been present for 20 years. The tumor appears encapsulated and contains large cystic spaces. There were prominent regressive changes which had led to bone formation. A. F. I. P. Acc. Nos. 219023–51 and 219023–52.

Figure 30. Photomicrograph of a typical neurilemoma taken from the tumor illustrated in figures 28 and 29. Antoni type A with palisading and Antoni type B with cystic spaces are illustrated. × 400. A. F. I. P. Acc. No. 219023–107.

*Case courtesy of Dr. Ira Gore, Henry Ford Hospital, Detroit, Mich.

Fig. 28

Fig. 29

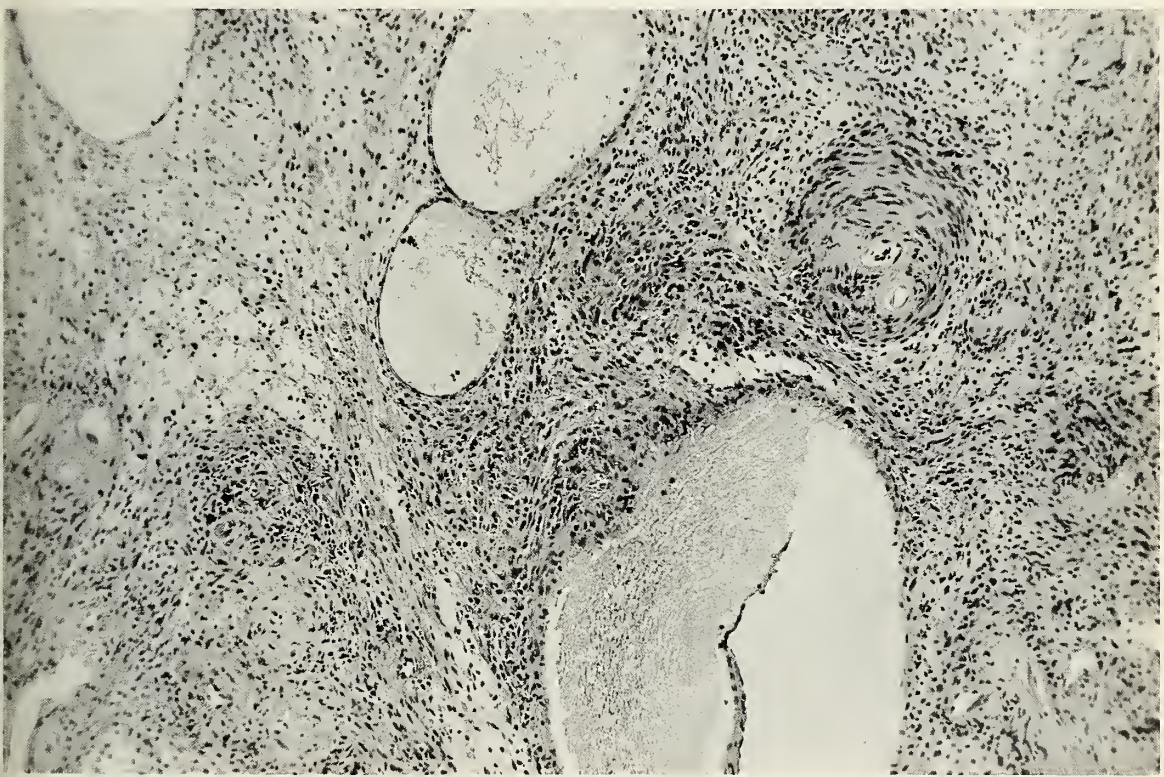

Fig. 30

MALIGNANT SCHWANNOMA

(Figures 31 and 32 are from the same case)

Figure 31. Roentgenogram showing partial calcification of the mass. A. F. I. P. Acc. No. 219023–54.

Figure 32. Gross photograph, demonstrating a large lobulated mass, grayish white on cut section. It was found at operation that the tumor involved the sacral plexus. Tumor was incompletely removed, and the patient died several years after operation. A. F. I. P. Acc. No. 219023–55.

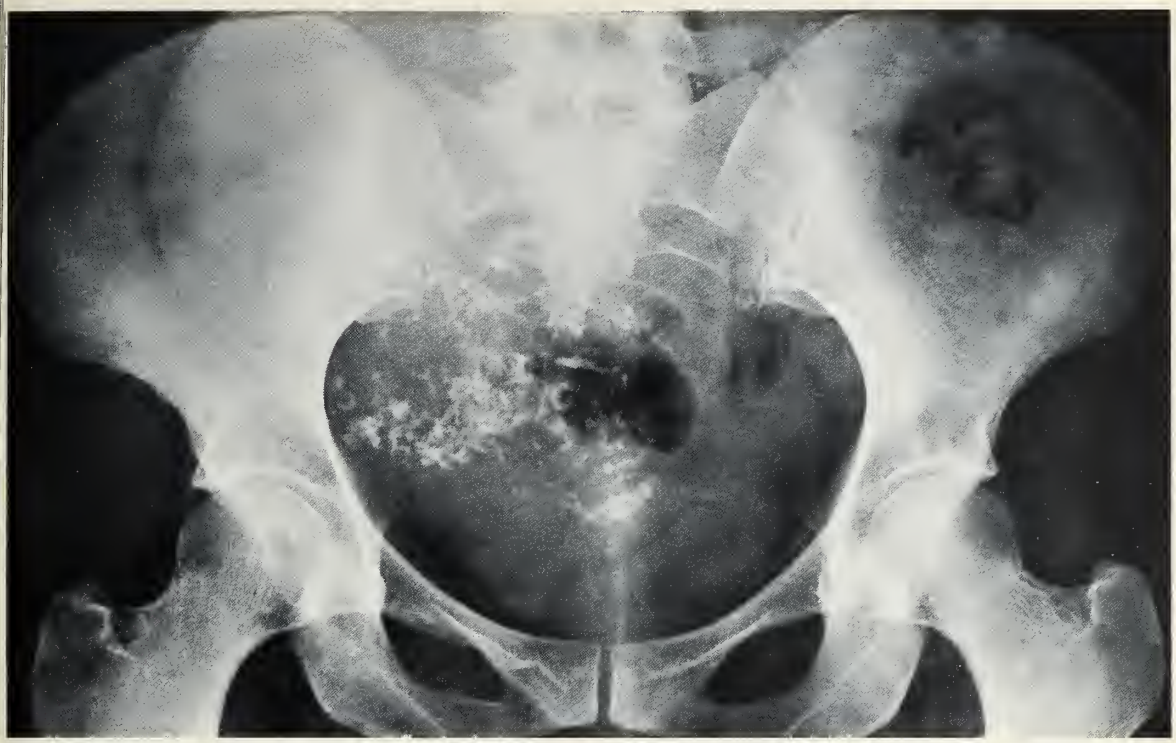

Fig. 31

Fig. 32

MALIGNANT SCHWANNOMA

(Figures 33 and 34 are from the same case)

Figure 33. Photomicrograph of another malignant schwannoma, showing well differentiated spindle-shaped cells with well defined nuclei and only rare mitotic figures. × 600. A. F: I. P. Acc. No. 219023–56.

Figure 34. The Wilder reticulin stain demonstrates reticulin in long wiry bundles, running in straight lines between cells rather than around them. × 600. A. F. I. P. Acc. No. 219023–57.

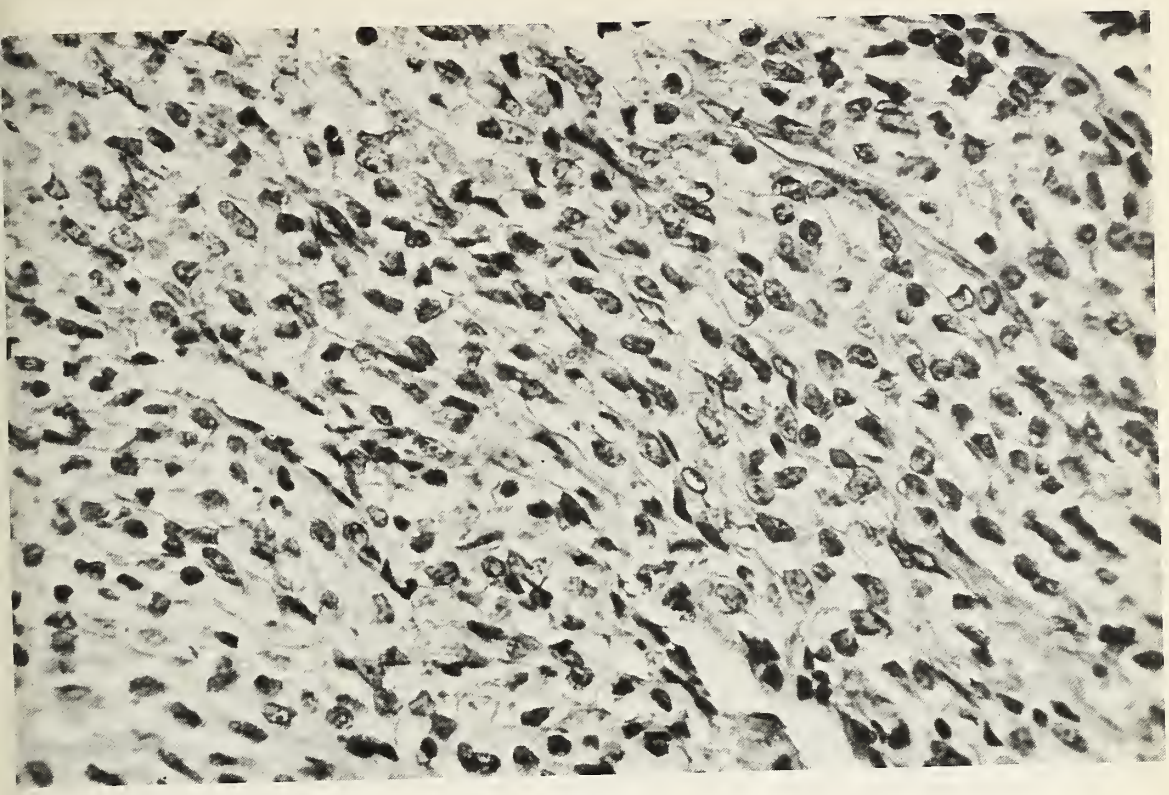

Fig. 33

Fig. 34

GANGLIONEUROMA

Figure 35. Well encapsulated ganglioneuroma from a 20-year-old woman. It measured 10 x 6 x 3 cm. and was attached to the sympathetic trunk from the first to fourth lumbar ganglia. (Courtesy of Dr. M. B. Dockerty, Mayo Clinic, Rochester, Minn.) A. F. I. P. Acc. No. 219023–58.

Figure 36. Differentiated retroperitoneal ganglioneuroma. Note ganglion cells and neurofibromatous stroma containing many neurites sheathed by Schwann cells. × 450. (From the Armed Forces Institute of Pathology.) A. F. I. P. Acc. No. 70650.

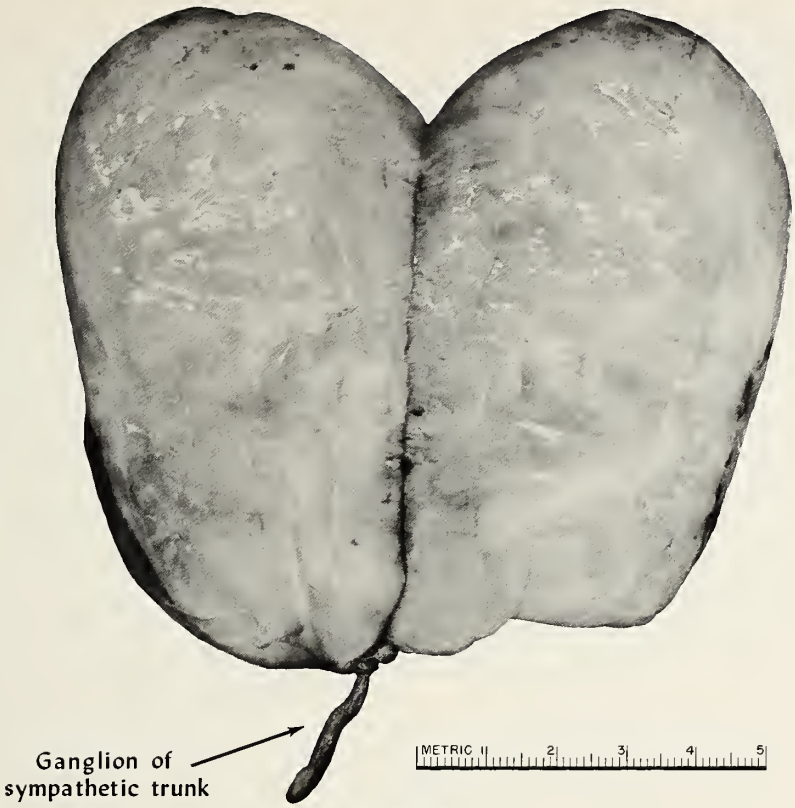

Fig. 35

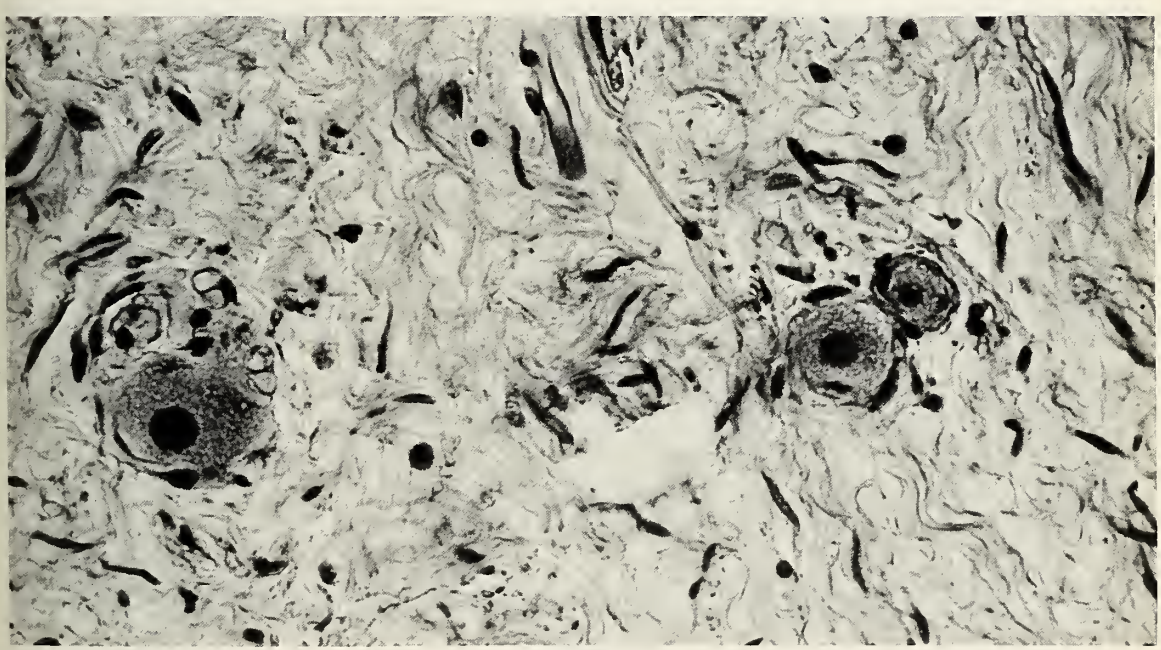

Fig. 36

GANGLIONEUROBLASTOMA

(Figures 38–40 are from the same case*)

Figure 37. Encapsulated retroperitoneal ganglioneuroblastoma with areas of hemorrhage. (Courtesy of Dr. M. B. Dockerty, Mayo Clinic, Rochester, Minn.) A. F. I. P. Acc. No. 219023–60.

Figure 38. Schematic drawing of the location of the tumor shown in figure 39. A. F. I. P. Acc. No. 219023–7.

*Case contributed by Dr. Charles Lockhard, Springfield, Mo.

METRIC II
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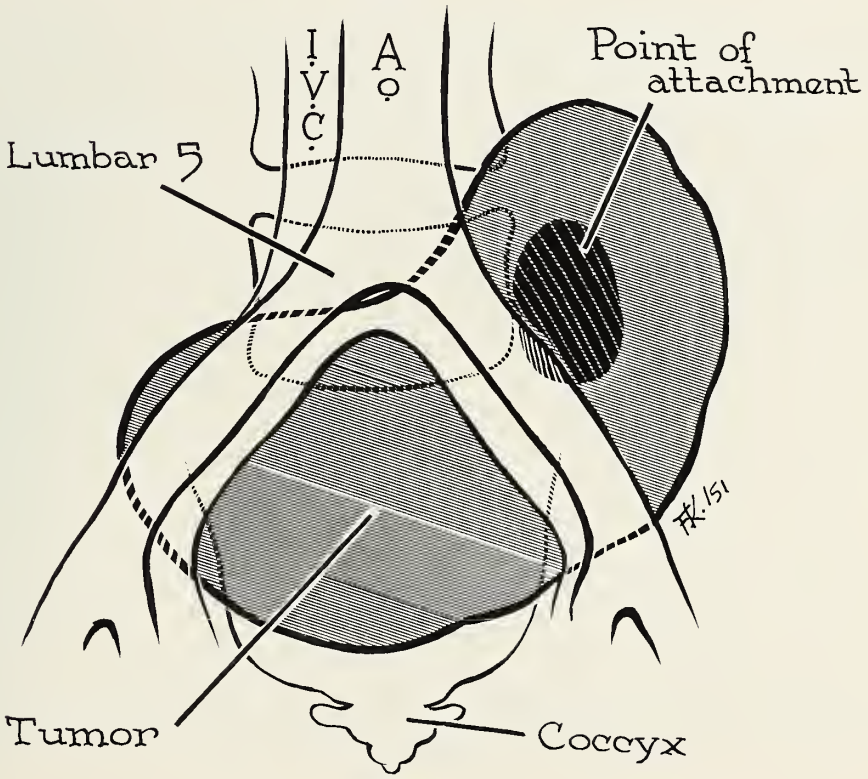
Fig. 37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V
C
A
Point of attachment
Lumbar 5
Tumor
Coccyx
FK. 151

Fig. 38

GANGLIONEUROBLASTOMA

(Figures 38–40 are from the same case*)

Figure 39. Artist's drawing of the relatively differentiated tumor of the sympathetic nervous system depicted in figure 40. A. F. I. P. Acc. No. 219023–62.

Figure 40. Photomicrograph of a ganglioneuroblastoma. The cells are larger than those seen in a neuroblastoma and are separated by feathery, cobwebby material. × 600. A. F. I. P. Acc. No. 219023–108.

*Case contributed by Dr. Charles Lockhard, Springfield, Mo.

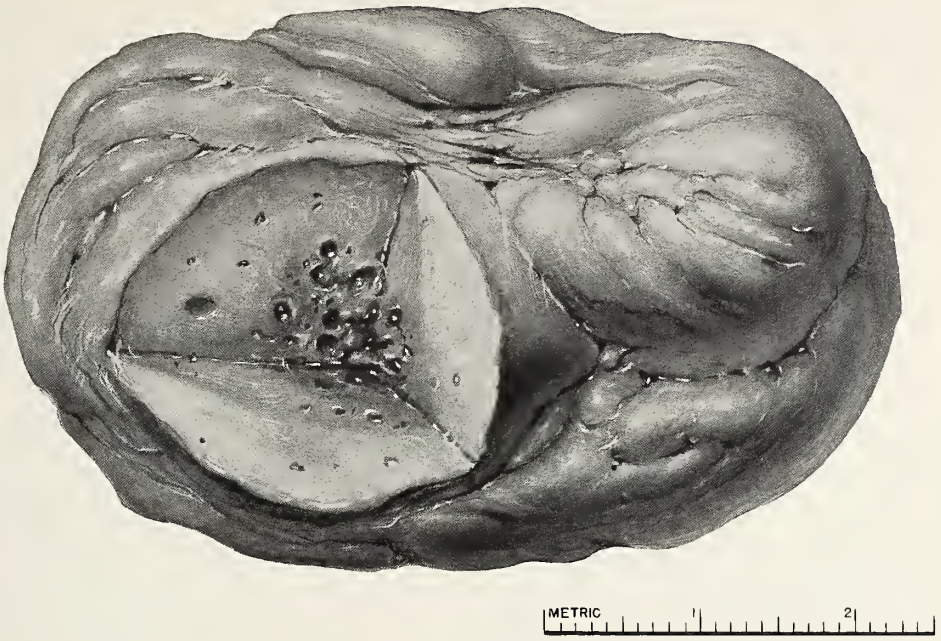

Fig. 39

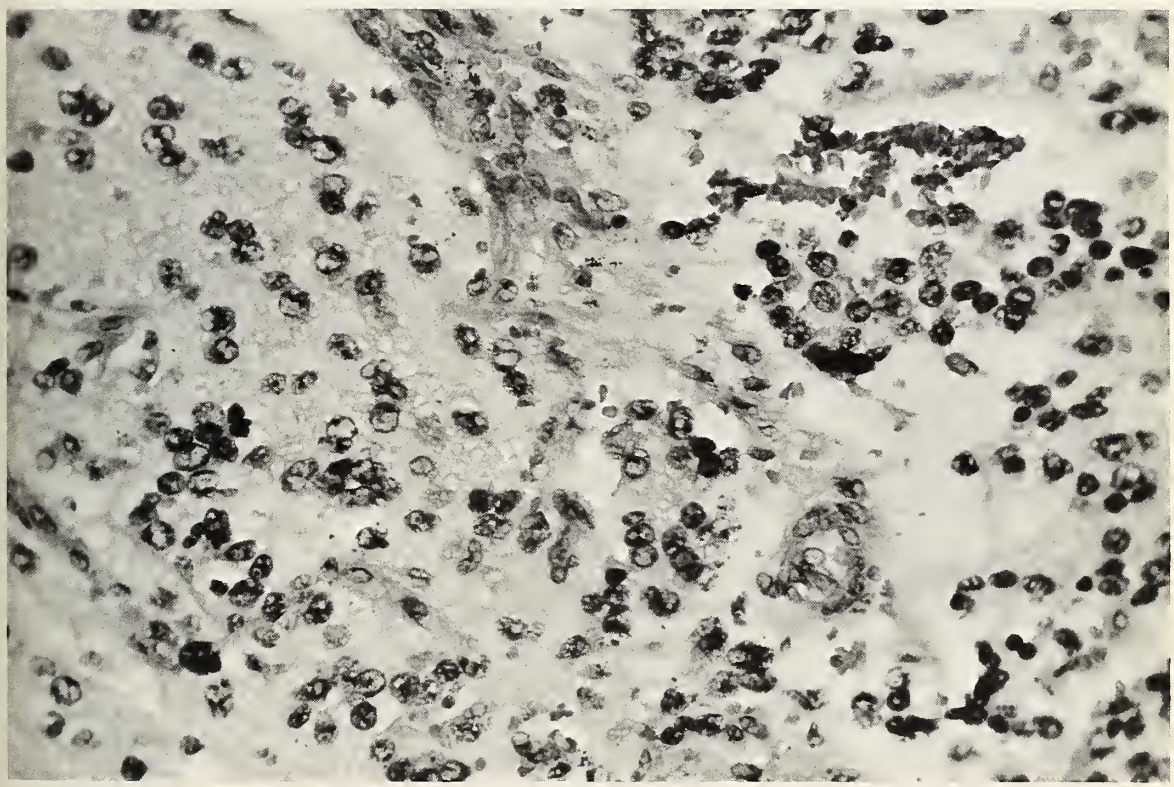

Fig. 40

neuroblastoma of the adrenal with metastases to the liver has been cured by irradiation (Wittenborg).

TUMORS ARISING FROM HETEROTOPIC CORTICAL ADRENAL TISSUE

Heterotopic adrenal cortical tissue has been reported in numerous locations; it is extremely common in the region of the adrenal, but has been found within the kidney substance, and is fairly frequently seen along the course of the spermatic and ovarian veins. In the region of the ovary it is usually found in the broad ligament, and in the region of the testis it is most prominent close to the tail of the epididymis. It is seldom found within the testis (Nelson) or within the ovary. Stout (personal communication) has seen heterotopic adrenal tissue in the canal of Nuck, in hernial and hydrocele sacs, in the mesentery of the appendix, and in the retroperitoneum between the two adrenal glands. It is questionable whether heterotopic adrenal tissue occurs in the liver. It is from the heterotopic cortical adrenal tissue which lies usually between the two adrenal glands retroperitoneally that tumors of cortical origin arise. Undoubtedly all the types described by Karsner in Fascicle 29, "Tumors of the Adrenal," could conceivably arise here, including **carcinomas of heterotopic cortical tissue.** We have seen two examples of nonfunctioning large **carcinomas arising from the adrenal cortex** (pl. II–B). These tumors were soft, cellular, and vascular, and had a poorly defined capsule. Microscopically the individual cells resemble cortical cells of the adrenal but often have bizarre nuclei (fig. 41).

TUMORS ARISING FROM FUNCTIONING AND NONFUNCTIONING PARAGANGLIONIC TISSUE

Tumors arising from functioning and nonfunctioning paraganglionic tissue have been observed: about 10 such cases have been reported. The functioning tumors with the clinical syndrome of sustained or intermittent hypertension are rare, but when they do appear are usually in the immediate region of one adrenal; in Biskind, Meyer, and Beadner's review of 29 cases, four were in this area. Cahill also reported a typical case of pheochromocytoma with sustained hypertension, in which a tumor was found at the bifurcation of the aorta, apparently arising from Zuckerkandl's body. In one case there was a second tumor in the epigastric area; postmortem examination confirmed the presence of two pheochromocytomas. Ganem and Cahill reported a case in which there were two pheochromocytomas, one in the adrenal gland and one in the retroperitoneal space, and Cragg described concurrent tumors of the left carotid body and of both Zuckerkandl's bodies. Those tumors not located in the adrenal can at times be discovered and diagnosed by their clinical syndrome or through the use of chemical tests (Calkins and Howard). Grossly and microscopically they resemble the pheochromocytomas found in the adrenal, but

ADRENAL CORTICAL CARCINOMA

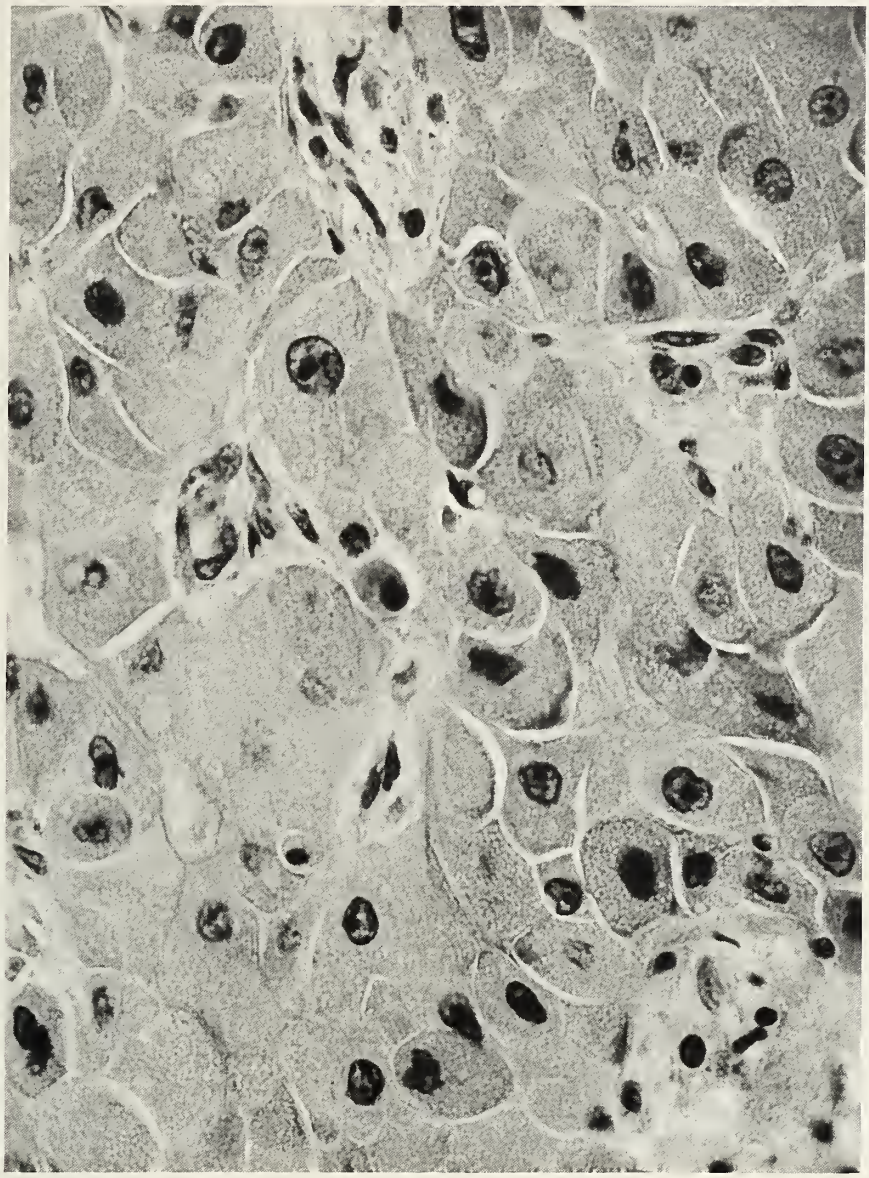

Figure 41. Well differentiated cortical carcinoma, showing variations in the cell and nuclear size and well defined cytoplasmic outlines. × 650. (Case contributed to the Armed Forces Institute of Pathology by Dr. A. S. Koenig, Fort Smith, Ark.) A. F. I. P. Acc. No. 235765.

PARAGANGLIOMA

Figure 42.* Nonfunctioning retroperitoneal encapsulated hemorrhagic paraganglioma arising from Zuckerkandl's body. A. F. I. P. Acc. No. 270229.

Figure 43.* Well differentiated paraganglioma. Paraganglion cells are arranged in small masses separated by capillaries. × 650. A. F. I. P. Acc. No. 331533.

*From the Armed Forces Institute of Pathology.

Fig. 42

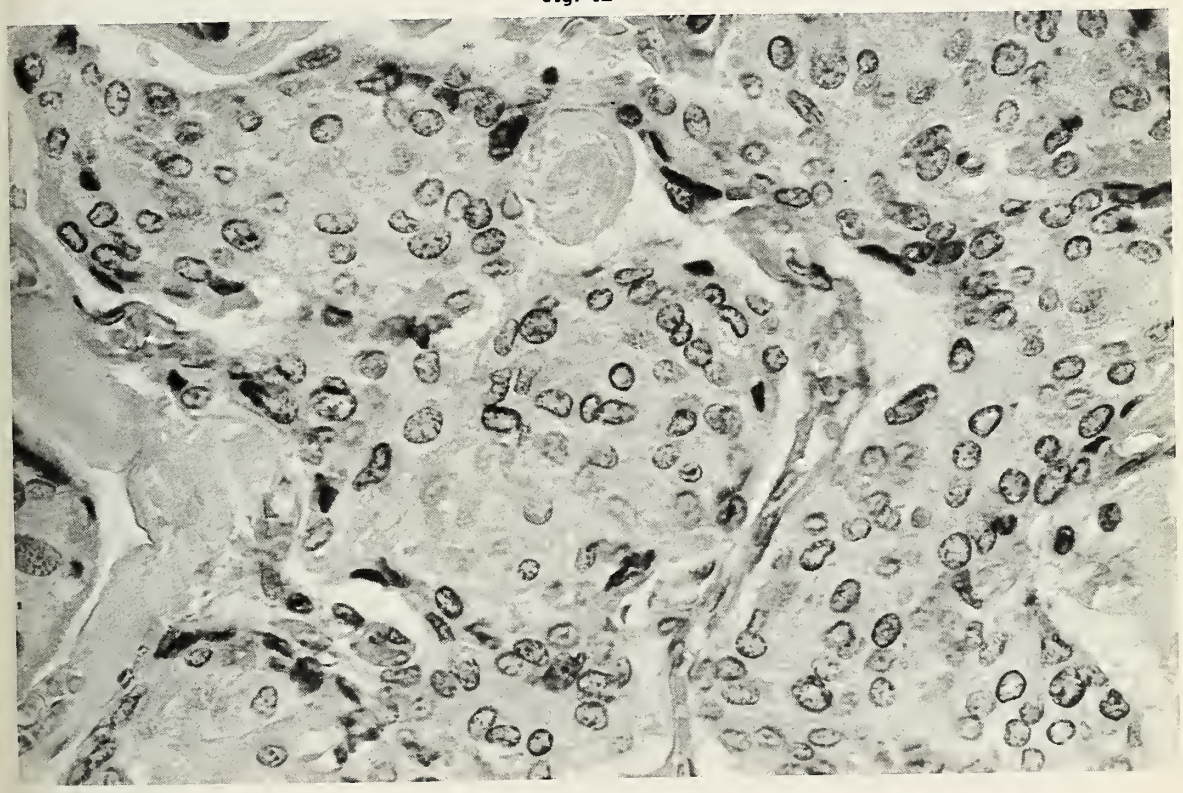

Fig. 43

PLATE III

PHEOCHROMOCYTOMA

Note the hemorrhagic character and encapsulation. This is a surgical specimen from a 70-year-old Negress who had had abdominal discomfort for many years. Her blood pressure ranged from 110/70 to 180/30. Two months prior to admission examination in the clinic revealed a firm left upper quadrant mass measuring 8 cm. Massage of the mass caused headache, increased perspiration, and slowing of the pulse. The blood pressure measured 300/160 and in five minutes it had returned to 122/74. Exploration showed a 6 cm. mass beneath the third portion of the duodenum. It was retroperitoneal, smooth, and ovoid. Her postoperative course was stormy but she has completely recovered. A. F. I. P. Acc. No. 219023–102.

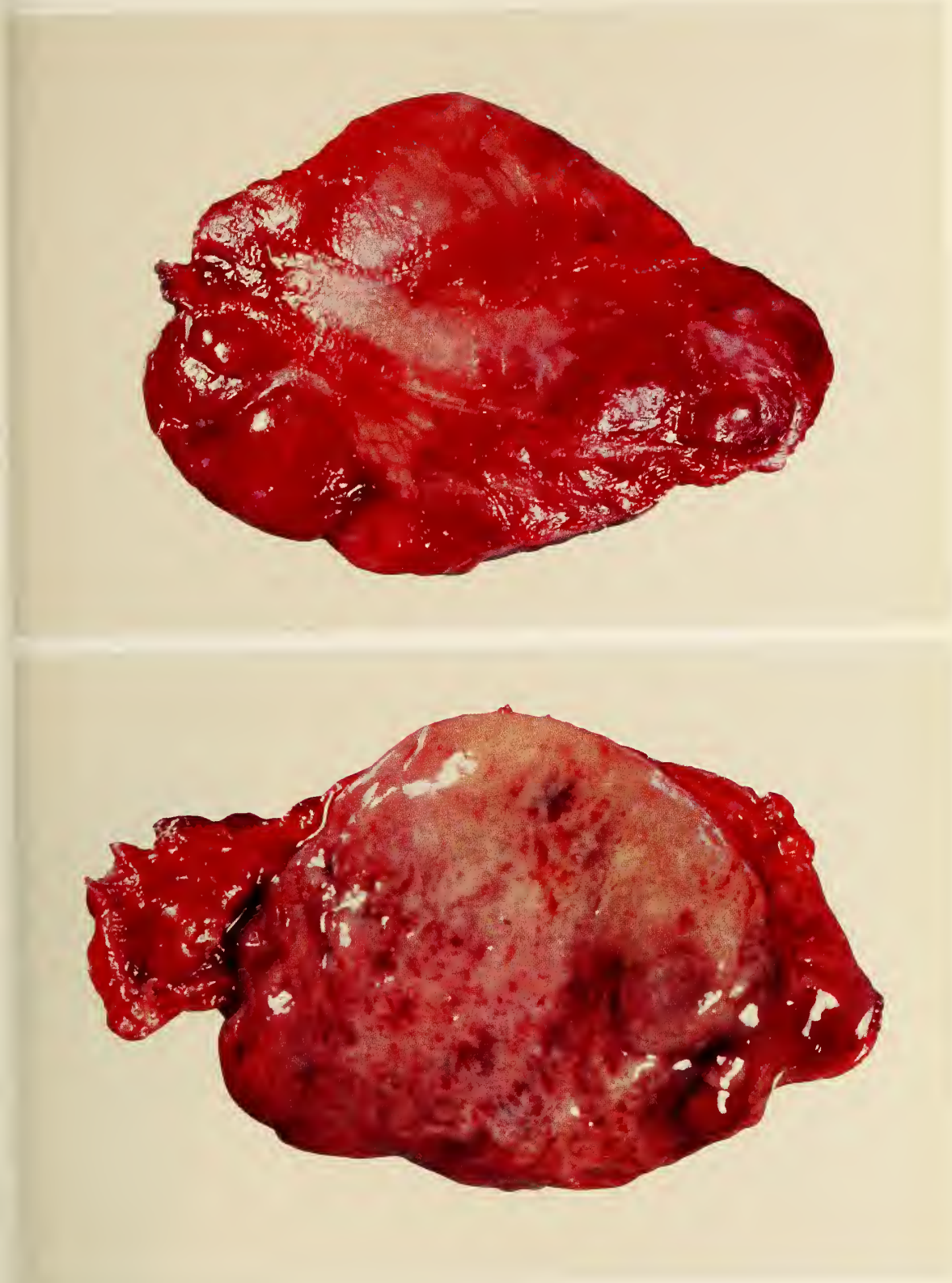

PLATE III

they may grow to a large size. Grossly they are usually benign, encapsulated, grayish red, highly vascular, and consequently difficult to remove (fig. 42, pl.III). Microscopically they have a typical appearance (fig. 43).

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ALVEOLAR SOFT PART SARCOMA

(MALIGNANT GRANULAR CELL MYOBLASTOMA, MALIGNANT NONCHROMAFFIN PARAGANGLIOMA)

Figure 44.* Cross section through a tumor which was adherent to a psoas muscle. Tumor was irregularly lobulated. A. F. I. P. Acc. No. 163529.

Figure 45.* Photomicrograph of malignant nonchromaffin paraganglioma (Smetana) demonstrating the general architecture of the tumor. This tumor shows groups of large cells separated by thin-walled capillaries. × 250. A. F. I. P. Acc. No. 120957.

Figure 46.* Higher magnification of nonchromaffin paraganglioma (Smetana). This demonstrates the relation of the tumor cells to capillaries. It also shows the well defined nuclei, prominent nucleoli, and absence of mitotic figures. × 1090. A. F. I. P. Acc. No. 262433.

*From the Armed Forces Institute of Pathology. These illustrations also appear in Smetana, H. F., and Scott, W. F. Malignant tumors of nonchromaffin paraganglia. Mil. Surgeon, 109: 330–349, 1951. Figures 1, 2, and 11 are our figures 44–46.

Fig. 44

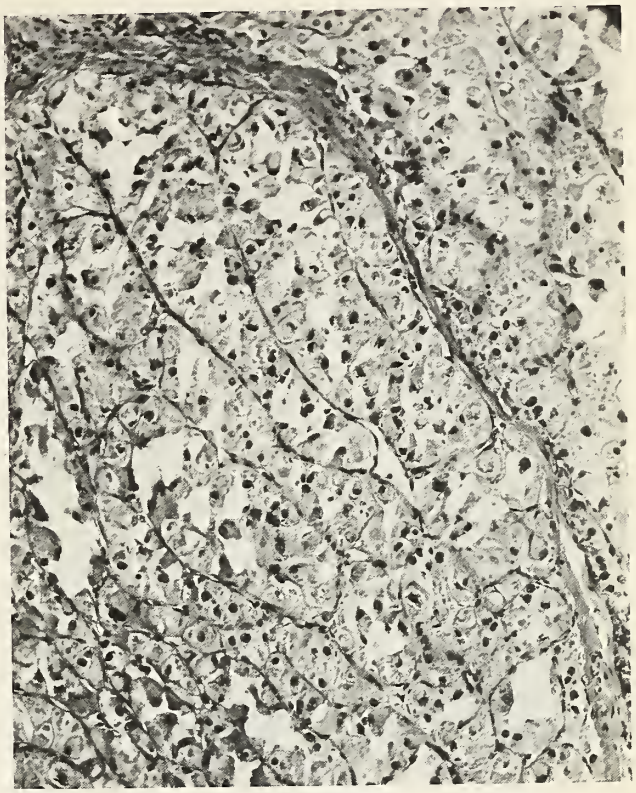

Fig. 45

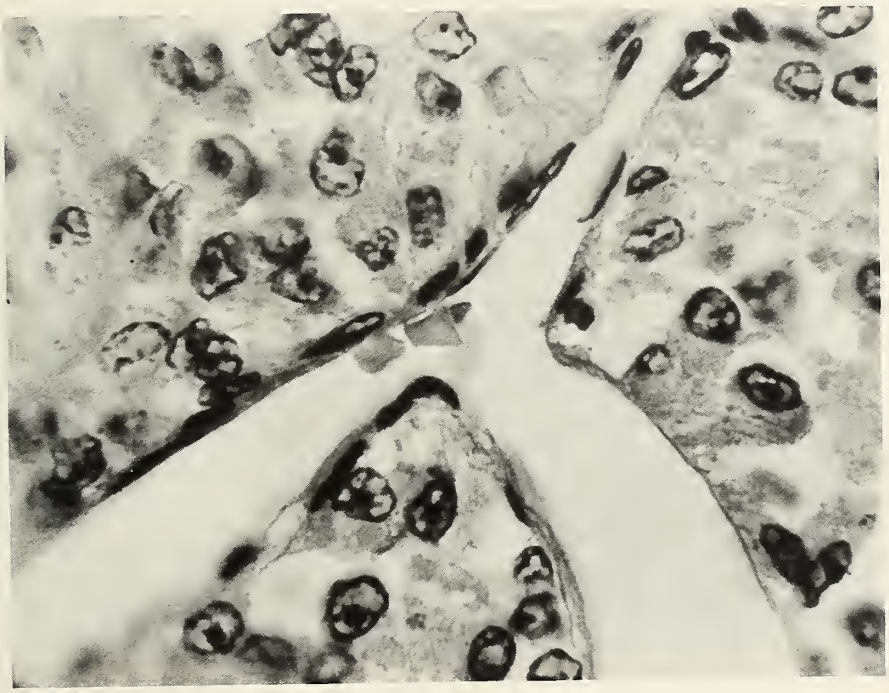

Fig. 46

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MISCELLANEOUS TUMORS

ALVEOLAR SOFT PART SARCOMA (malignant nonchromaffin paraganglioma, malignant granular cell myoblastoma)

The histogenesis of this rare malignant neoplasm is still in doubt. In the past this tumor has been called a malignant granular cell myoblastoma (Ravich, Stout, and Ravich; Ackerman and Phelps). Murray cultured in vitro two benign granular cell myoblastomas and one tumor of the malignant granular cell type. She thought that the cultural characteristics of all three of these neoplasms bore a greater resemblance to cultures of various forms of skeletal muscle, normal and neoplastic, than to cultures of other tissue types to which their origin has sometimes been attributed. Christopherson, Foote, and Stewart prefer to call this neoplasm by the noncommittal term "alveolar soft-part sarcomas," but do state that "the histological resemblance to paragangliomas is impressive." Both Stout and Stewart point out that the carotid body tumors rarely metastasize while this neoplasm often does. Smetana and Scott are of the opinion that the tumor should be called a malignant nonchromaffin paraganglioma, basing their theory of histogenesis on the similarity of its pattern to the carotid body tumor; emphasizing this resemblance are the masses of well differentiated cells of organoid character separated by capillaries. This pattern is well brought out by a reticulin stain. This tumor frequently occurs in the thigh, and Johnson has demonstrated paraganglionic glomera in the soft tissue surrounding the femoral vessels. It also occurs in the retroperitoneal region; Smetana and Scott have described four such cases. Grossly the tumors are pseudoencapsulated, and on cross section are highly vascular and are divided into lobules and separated by delicate connective tissue septa (fig. 44). The microscopic diagnosis is made on the basis of the typical organoid arrangement with groups of cells separated by capillaries (fig. 45) and on further delineation by the reticulin stain. Individual tumor cells are rather large and often have a granular cytoplasm; the nuclei are well defined and have prominent nucleoli (fig. 46). Mitotic figures are not present. Frequently tumor cells may be seen within blood vessels. This tumor is often mistaken for a carcinoma of the kidney, an undifferentiated carcinoma, a liposarcoma, or even a chordoma. Unlike the liposarcoma, the viable tumor cells do not show cytoplasmic fat by ordinary fat stains. No matter what the histogenesis, this tumor is a primary malignant neoplasm that frequently recurs locally and may have distant metastases.

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REMNANTS OF RENAL BLASTEMA

Hansmann and Budd studied 17 cases of unattached retroperitoneal tumors which they thought arose from remnants of the embryonal urogenital apparatus; they divided these tumors into solid and cystic types. In a review of the literature they found 37 other cases. Many of these tumors resembled cystadenomas of the ovary, and others were malignant with a suggestion of origin from renal tubules. There appears little doubt that such neoplasms exist, but they form only a small fraction of the total group of malignant retroperitoneal tumors. Their identification, however, is often by exclusion, and their recognition is based on rather uncertain histologic criteria. The few cases which we have seen have been identified only tentatively because of their resemblance to renal tubules and because at times these cancer cells contain fat (fig. 47). Stewart has only rarely placed a tumor in this category; and in making the diagnosis, he wrote, "The suggestion was always on sections from outside sources usually difficult to document and not certain." Stout has not been able to submit any case about which he felt satisfied that the tumor arose from embryonic urogenital apparatus.

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TUMORS ARISING FROM EMBRYONIC REMNANTS

Retroperitoneal **benign teratomata** are relatively rare tumors, only about 200 having been reported in the literature: 40 of these were first observed in infants or little children (Arnheim; figs. 48–54). Most of them develop in females, and many patients with this type of tumor may be cured by surgery

(fig. 54). These tumors grow slowly, are often present at birth, and are frequently located between the rectum and sacrum. They may grow to a large size and cause partial obstruction to the rectum, and because of inflammation may become adherent to the surrounding structures; this adherence secondary to inflammation may lead the surgeon to believe that he is dealing with a malignant tumor. The tumor may occupy the complete retroperitoneal space on one or both sides, and is often found high in the retroperitoneal space. It may grow through the sacrosciatic foramen and present as a swelling of the buttock (Galletly). Death may occur because of pressure effects. Grossly these tumor lesions are cystic (fig. 51) and often contain well organized tissue with rudimentary organs (Nicholson). There may be segments of intestine with mesentery, bronchi with cartilaginous rings, and imperfect extremities. Microscopically such neoplasms contain every conceivable type of tissue, including thyroid, adrenal, respiratory epithelium, lung, sympathetic ganglia, glial tissue, salivary gland, smooth and striated muscle, fat, fibrous tissue, skin appendages, etc. (figs. 52, 53). Infrequently these tumors may undergo malignant change followed by rapid invasion of surrounding structures and metastases (Stewart, Alter, and Craig; Ravitch and Smith; Gross, Clatworthy and Meeker; fig. 55). The microscopic types of **malignant teratomas** are extremely variable (figs. 56–58).

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TUMORS ARISING FROM NOTOCHORD

Sacrococcygeal **chordomas** arising from remnants of the fetal notochord involve the retroperitoneal space by direct extension. Ribbert demonstrated that by puncturing the nucleus pulposus in rabbits he produced a tumor identical with chordoma. There have been about 135 cases reported. They are

REMNANTS OF RENAL BLASTEMA

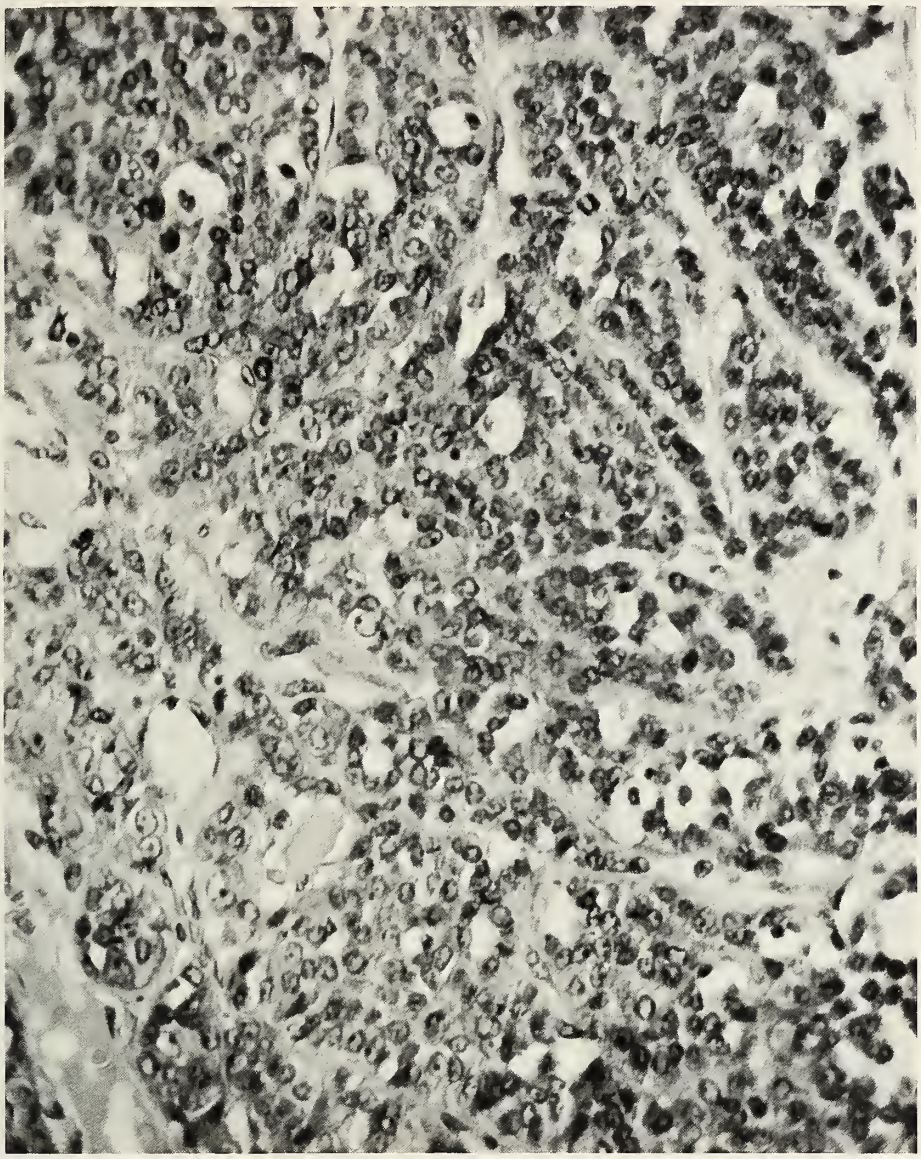


Figure 47. Photomicrograph of a large retroperitoneal tumor demonstrating a pattern suggestive but not diagnostic of renal anlage origin. × 395. A. F. I. P. Acc. No. 219023-22.

TERATOMA

Figure 48. Bisected infant showing extensive retroperitoneal teratoma. (Courtesy of Dr. Roswell K. Brown, Buffalo, N. Y.) A. F. I. P. Acc. No. 219023–65.

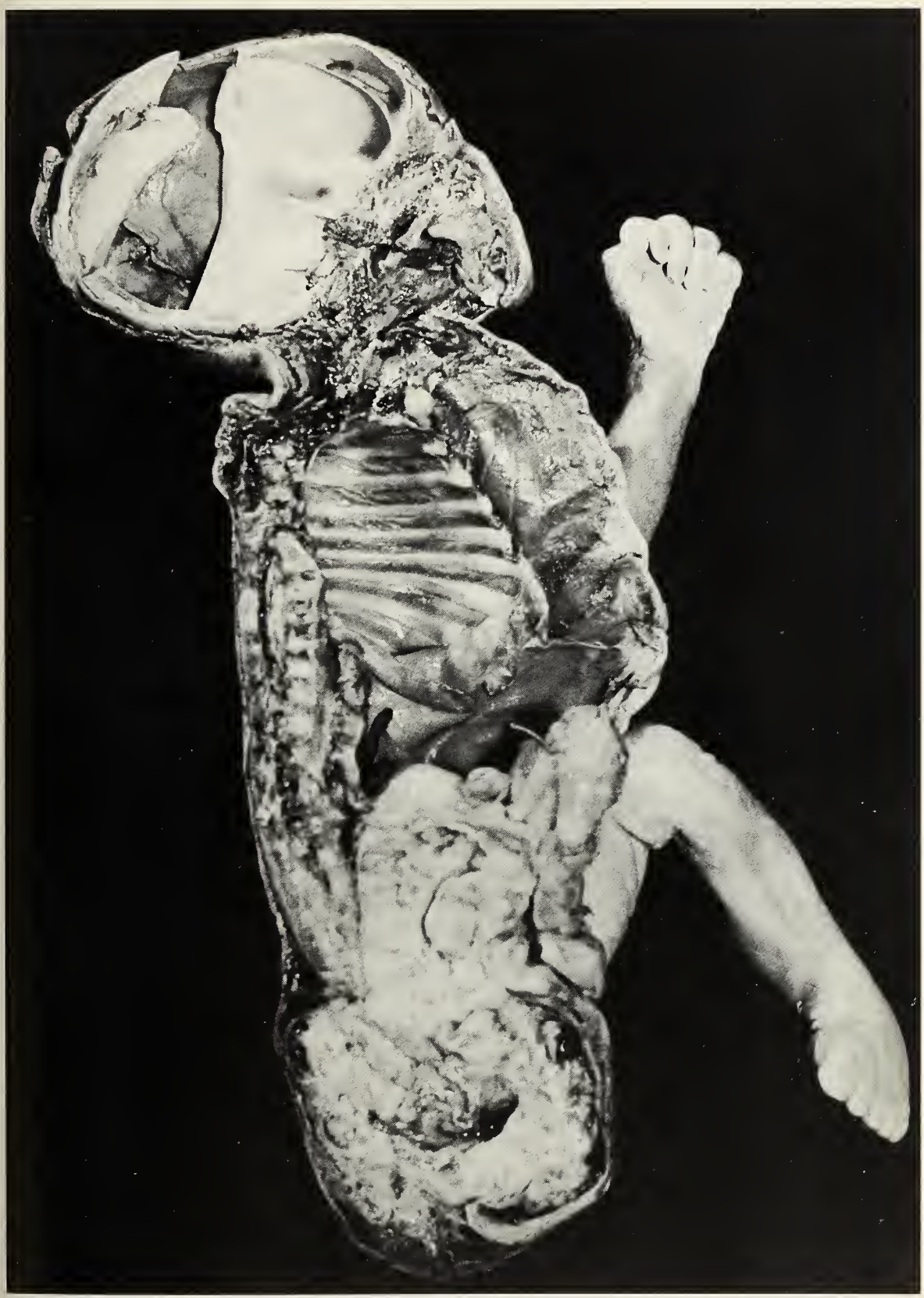


Fig. 48

TERATOMA

(Figures 49 and 50 are from the same case)

Figure 49. Clinical photograph of a 7-week-old child with a huge retroperitoneal benign teratoma. This was successfully removed, and the child is well. A. F. I. P. Acc. No. 219023–64.

Figure 50. Lateral film demonstrating the well delimited tumor mass. A. F. I. P. Acc. No. 219023–65.

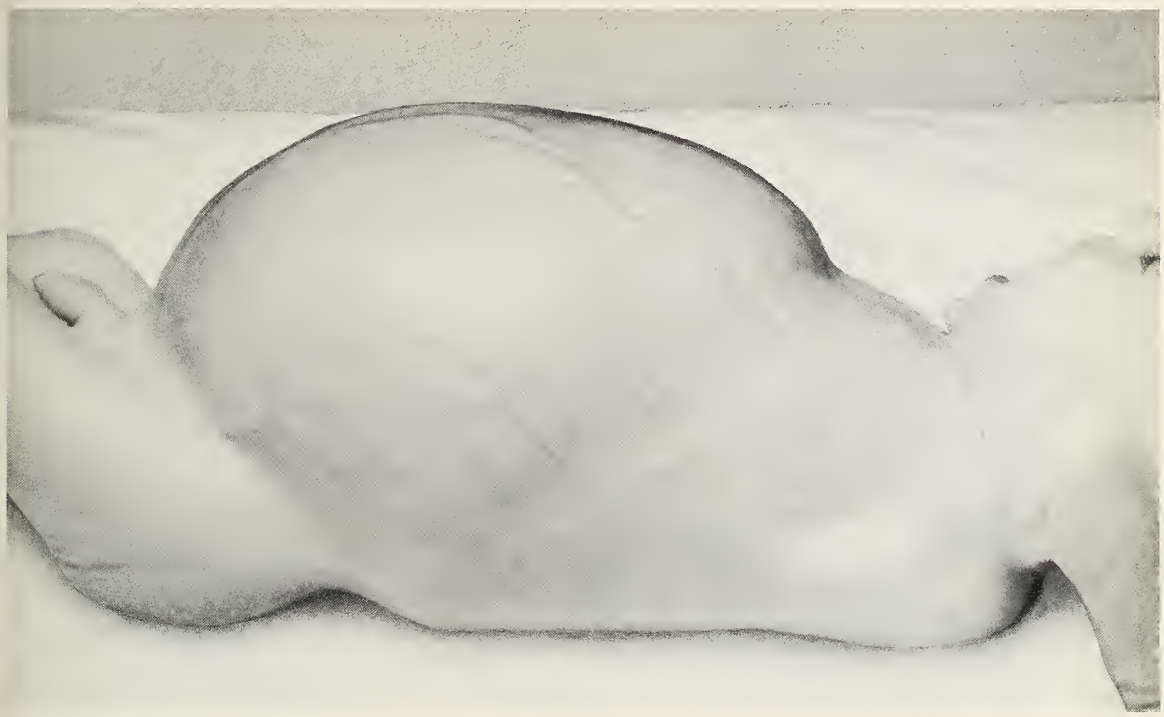


Fig. 49

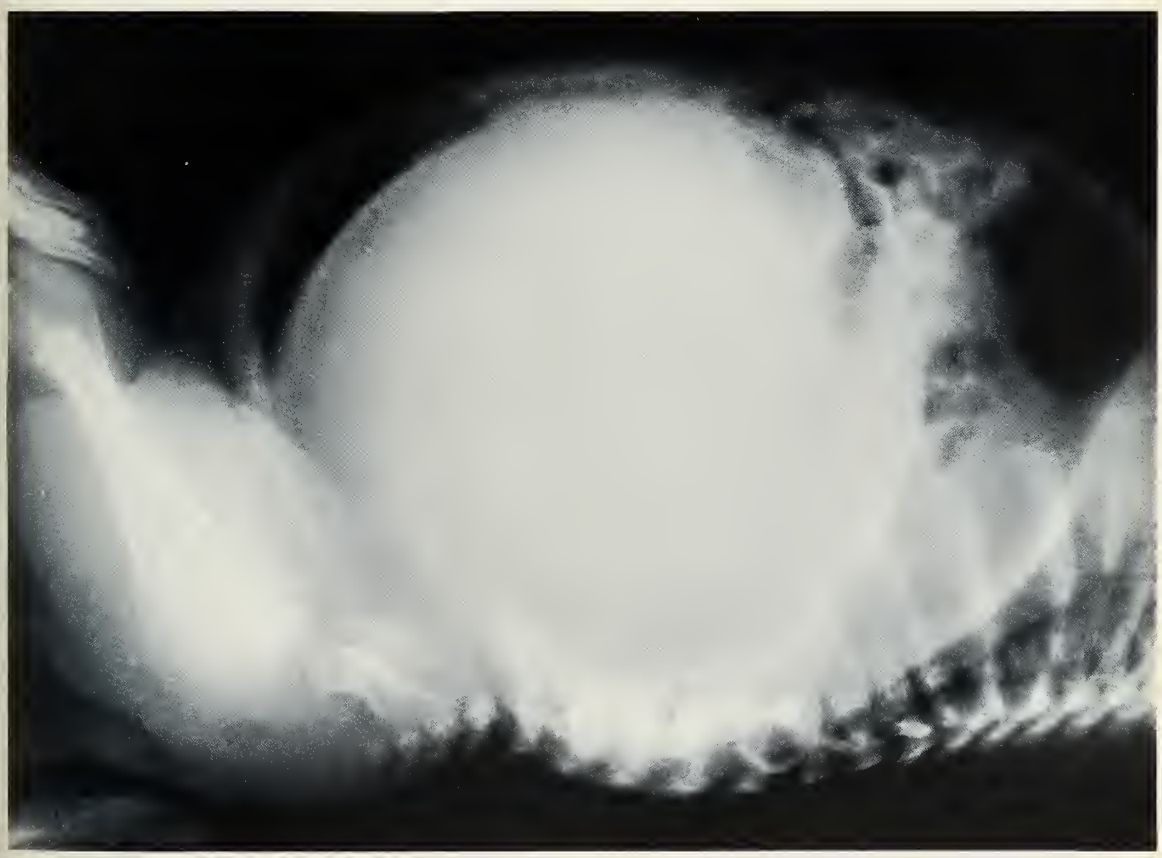


Fig. 50

TERATOMA

(Figures 51 and 52 are from the same case*)

Figure 51. Gross photograph of three vertical slides of a retroperitoneal teratoma obtained at postmortem in a 5-month-old female child. It measured 14 x 7 x 6 cm., weighed 160 gm., and contained a great variety of tissues. A. F. I. P. Acc. No. 219023–66.

Figure 52. Photomicrograph showing structures resembling choroid plexus. × 175. A. F. I. P. Acc. No. 219023–67.

Figure 53. Stratified squamous epithelium in a well differentiated retroperitoneal teratoma. × 210. (From the Armed Forces Institute of Pathology.) A. F. I. P. Acc. No. 147815–1.

*Courtesy of Dr. Rupert A. Willis, Leeds, England.

Fig. 51

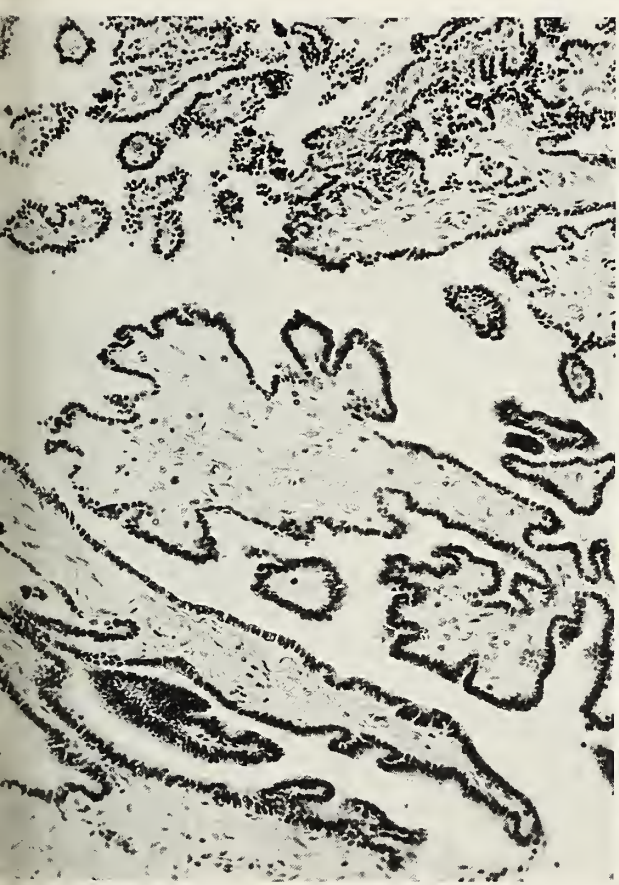


Fig. 52

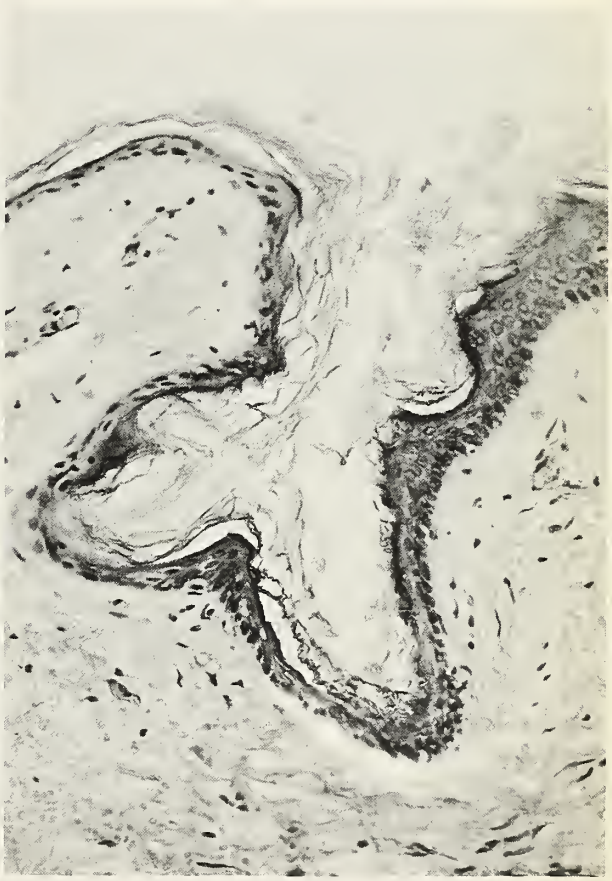


Fig. 53

TERATOMA

Figure 54. Gross photograph of the external and cut surface of an almost completely solid teratoma, weighing 600 gm. This tumor was removed surgically from a 5-day-old child by Dr. Leon Goldman, San Francisco, Calif. Seven years after removal, December 3, 1949, the child was living and well, without evidence of disease. (Courtesy of Dr. L. Goldman and Dr. H. Glen Bell, San Francisco, Calif.) A. F. I. P. Acc. No. 219023–69.

Figure 55. Malignant retroperitoneal teratoma occurring in a 6-year-old child. Specimen measured 15 cm. and weighed 350 gm. The spleen, seen at the upper border of the mass, and a part of the pancreas were removed with the tumor. Patient died of spread of the disease approximately five months after removal. (This is figure 12 in Arnheim, E. E. Retroperitoneal teratomas in infancy and childhood. Pediatrics, 8: 309–327, 1951.) A. F. I. P. Acc. No. 219023–68.

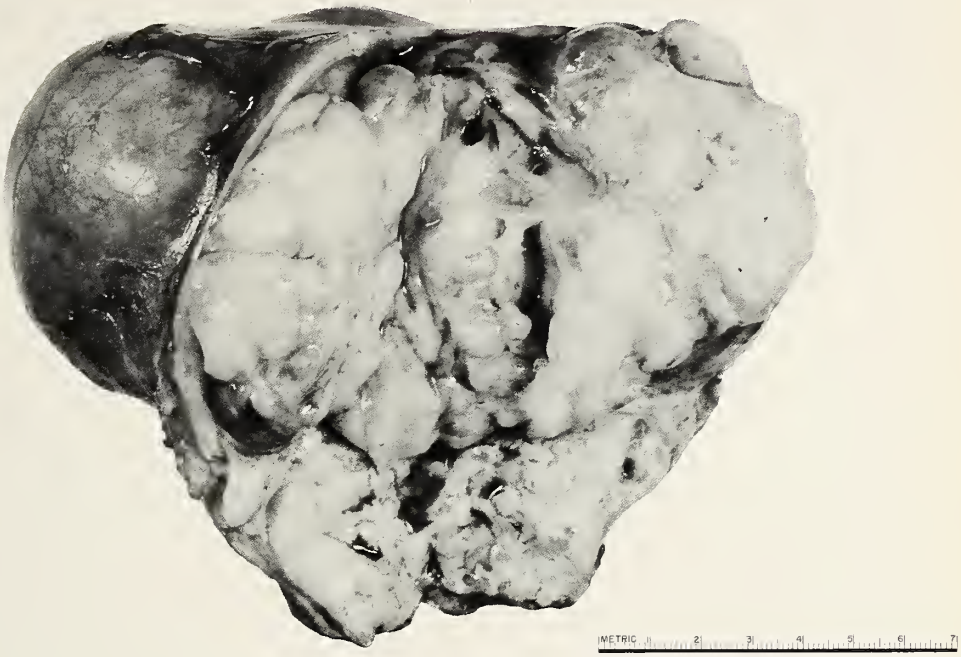


Fig. 54

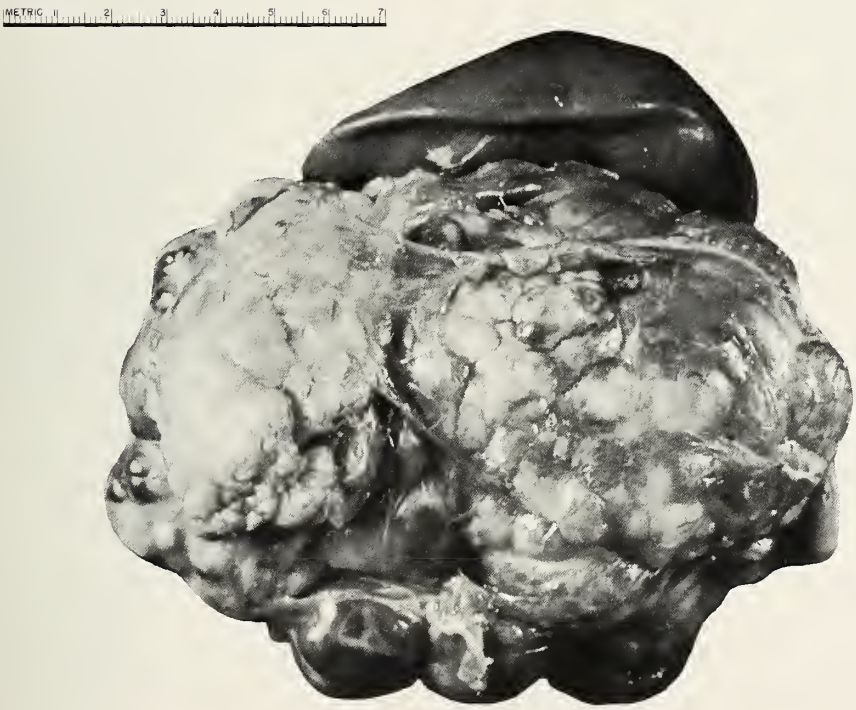


Fig. 55

MALIGNANT TERATOMA

(Figures 56–58 are from the same case*)

Figure 56. Squamous cells and malignant stroma in a malignant retroperitoneal teratoma. Note the great variation in the microscopic pattern of this and the two following photomicrographs. × 185. A. F. I. P. Acc. No. 104822–2.

Figure 57. Undifferentiated tumor cells. × 435. A. F. I. P. Acc. No. 104822–1.

Figure 58. Questionable respiratory epithelium amid undifferentiated tumor. × 310. A. F. I. P. Acc. No. 104822–3.

*From the Armed Forces Institute of Pathology.

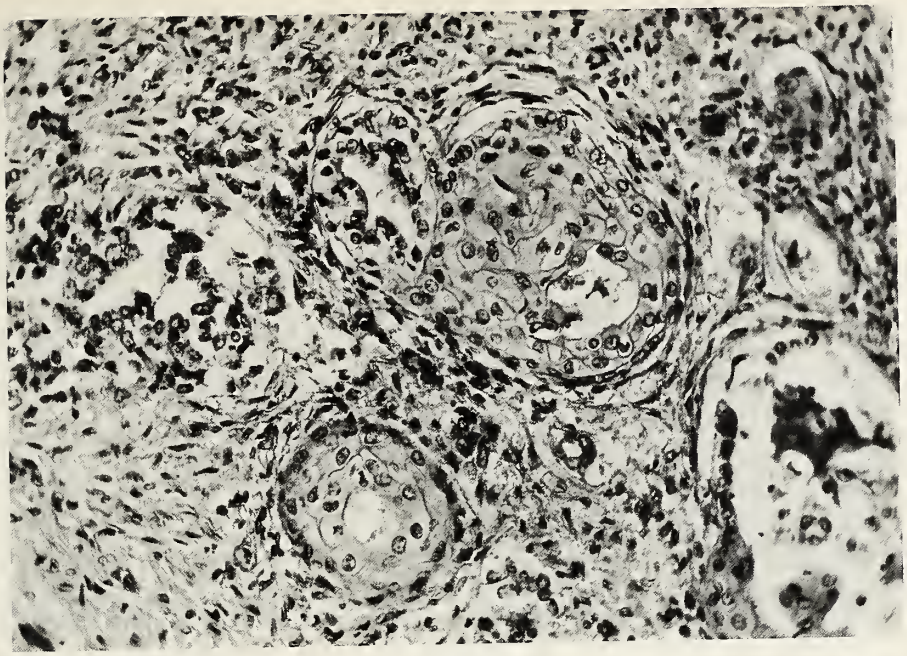


Fig. 56

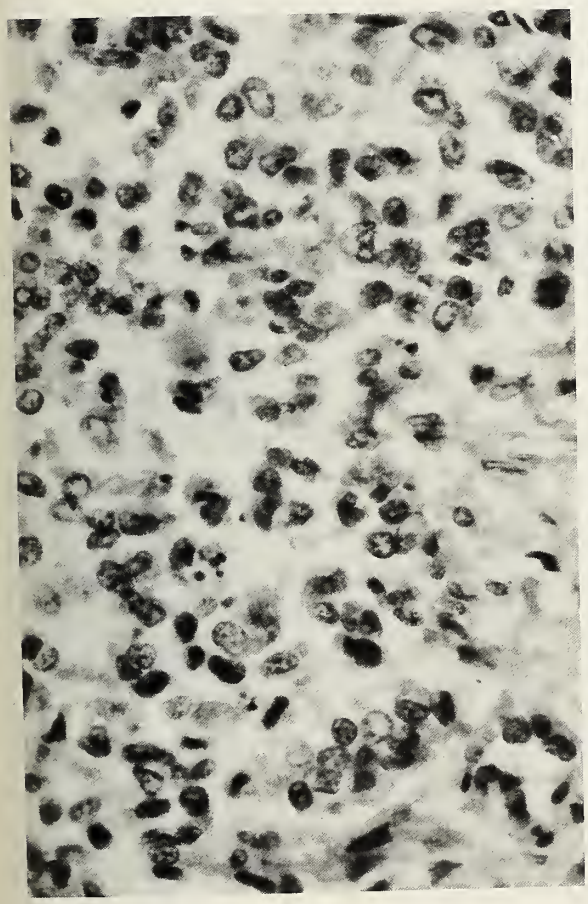


Fig. 57

Fig. 58

PLATE IV

CHORDOMA

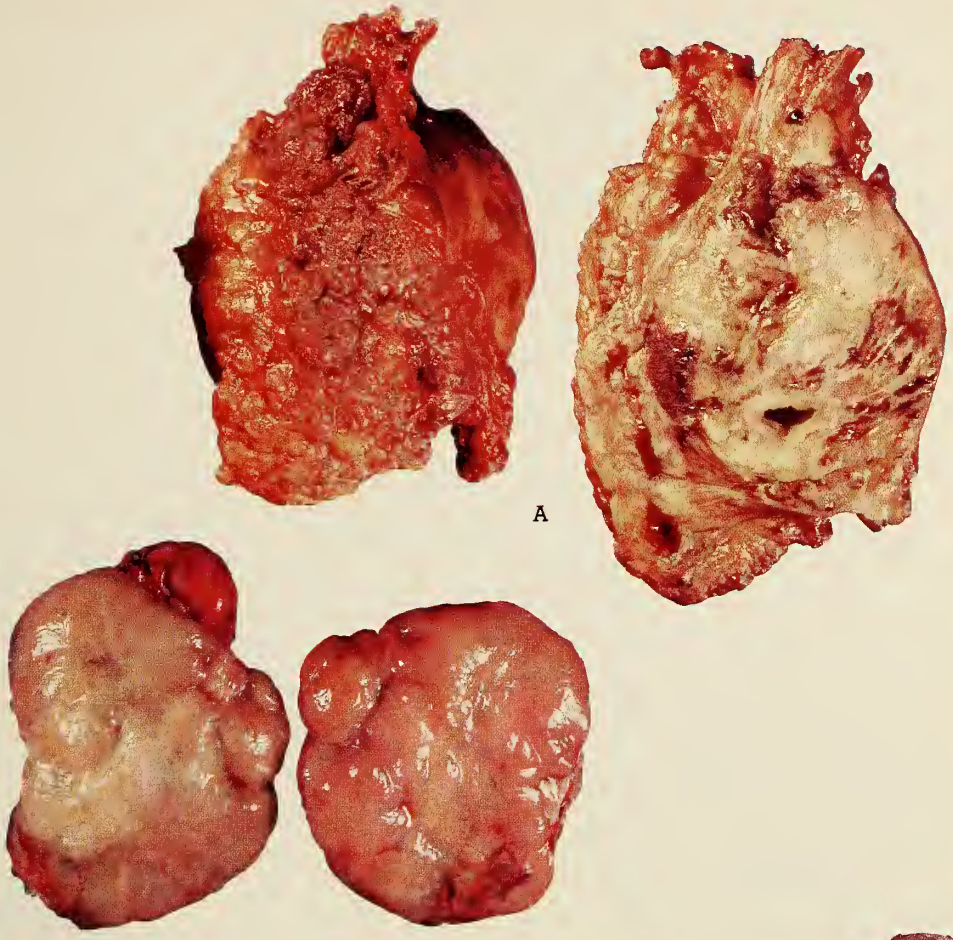
A. Specimen of a large chordoma measuring 11 x 8 x 7 cm., weighing 420 gm. It is hemorrhagic and somewhat gelatinous, with areas of firm grayish white tissue. This chordoma occurred in a 65-year-old woman. Symptoms were of two years' duration. The tumor was radically excised with a segment of the sacrum on July 11, 1950. A. F. I. P. Acc. No. 219023–25.

LIPOSARCOMA

B. Gross specimen of large homogeneous liposarcoma of the mesentery. A. F. I. P. Acc. No. 219023–C3. (Contributed by Dr. A. P. Stout, Columbia University, College of Physicians and Surgeons, New York, N. Y.)

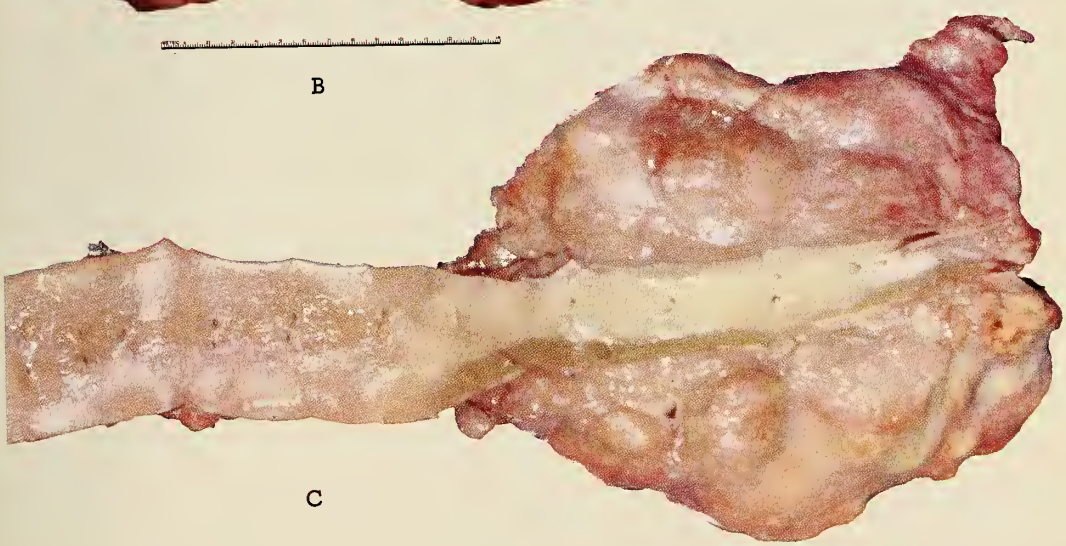
METASTATIC TUMOR

C. Extensive retroperitoneal metastases involving periaortic lymph nodes in a patient with a primary malignant testicular tumor. This distribution of lymph node involvement also occurs in the lymphomas, such as Hodgkin's disease and lymphosarcoma. A. F. I. P. Acc. No. 219023–C4.



A

B



C

PLATE IV

CHORDOMA

(Figures 59 and 60 are from the same case)

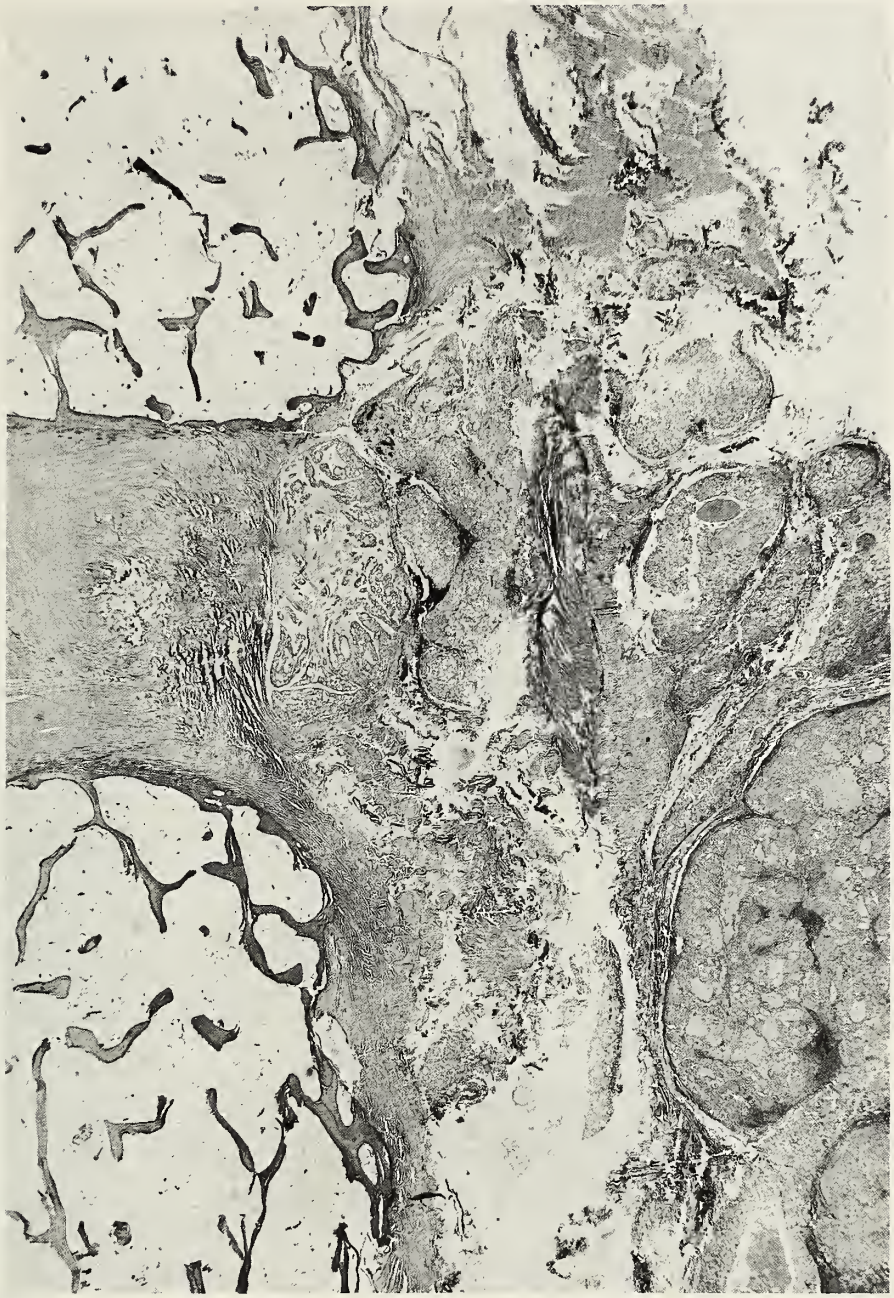


Figure 59. Photomicrograph of a chordoma arising in the curve of the sacrum and growing out from one of its intervertebral discs. The patient, a 43-year-old male physician, had had spasms of pain for many years. Roentgenogram showed no changes in the sacrum. The lesion was resected by block removal of a portion of the sacrum with the tumor. Low power. A. F. I. P. Acc. No. 219023–70.

most frequent in men, can occur at any age, but usually are found in the fifth decade (Gentil and Coley). The notochord develops from entoderm and in the embryo is a solid cord of tissue lying ventral to the neural canal. With further development of the embryo, mesodermal tissue surrounds it, and from this tissue intervertebral discs and vertebrae develop. Remnants of the notochord lie entirely within the vertebrae, in the intervertebral disc, and are rarely unattached in the soft tissues close to the sacrum. Most of the tumors arise from within the bone rather than from within the disc (Harvey and Dawson). Berard, Dunet, and Peyron demonstrated notochordal tissue lying on the surface of the vertebral bodies chiefly in the lumbosacral area. These tumors arise most frequently in the craniopharyngeal and sacrococcygeal zone, but are most common in the sacrococcygeal area. In this zone they grow slowly; the duration of symptoms before diagnosis is made is usually over five years (fig. 59). In its growth the tumor invariably destroys a portion of the sacrum usually by an osteolytic process (figs. 61, 62) or rarely is itself replaced by an osteoblastic process (Wood and Himadi). If the tumor encroaches on the spine, symptoms of a spinal cord tumor arise. The tumor then pushes its way into the retroperitoneal space, where it may grow large enough to narrow the lumen of the large bowel or even impinge upon the bladder. It can be felt as a firm extrarectal mass. These nonencapsulated tumors are soft and gelatinous, with areas of hemorrhage (pl. IV–A). In a few instances it may be possible to excise them radically, but in most instances this is impossible. We have had one instance in a 43-year-old physician who over a 5-year period had clinical symptoms of severe paroxysmal pain aggravated at the time of bowel movements. On rectal examination a small mass could be felt in the hollow of the sacrum. Radical excision was possible. This chordoma apparently arose from one of the intervertebral spaces of the sacrum (fig. 59). Irradiation therapy may give considerable palliation over long periods of time (figs. 61–64). Microscopically these tumors closely resemble notochordal tissue as observed in the nuclei pulposi of the intervertebral discs, and the neoplasms grow in cords (fig. 60). Alezais and Peyron believe that the various appearances of chordoma are consistent with the development of the primitive notochord. Individual cells range from small cells with small nuclei and fine nucleoli to extremely large cells (fig. 64). These large cells have a vacuolated cytoplasm and infrequently the nucleus also appears vacuolated. The largest vacuolated cells have been designated as physaliferous (physallis, a bubble). Some of these vacuoles contain glycogen (Stewart and Morin). Fine fibrils appear in the vacuolated zone, and at times the cells may have a compressed fusiform shape. Mitotic figures are not present. The neoplasm may have extremely confusing areas of cartilage formation (Mabrey). In a few instances these tumors have metastasized regionally or distantly (Morris and Rabinovitch). If the chordoma shows

CHORDOMA

(Figures 59 and 60 are from the same case)

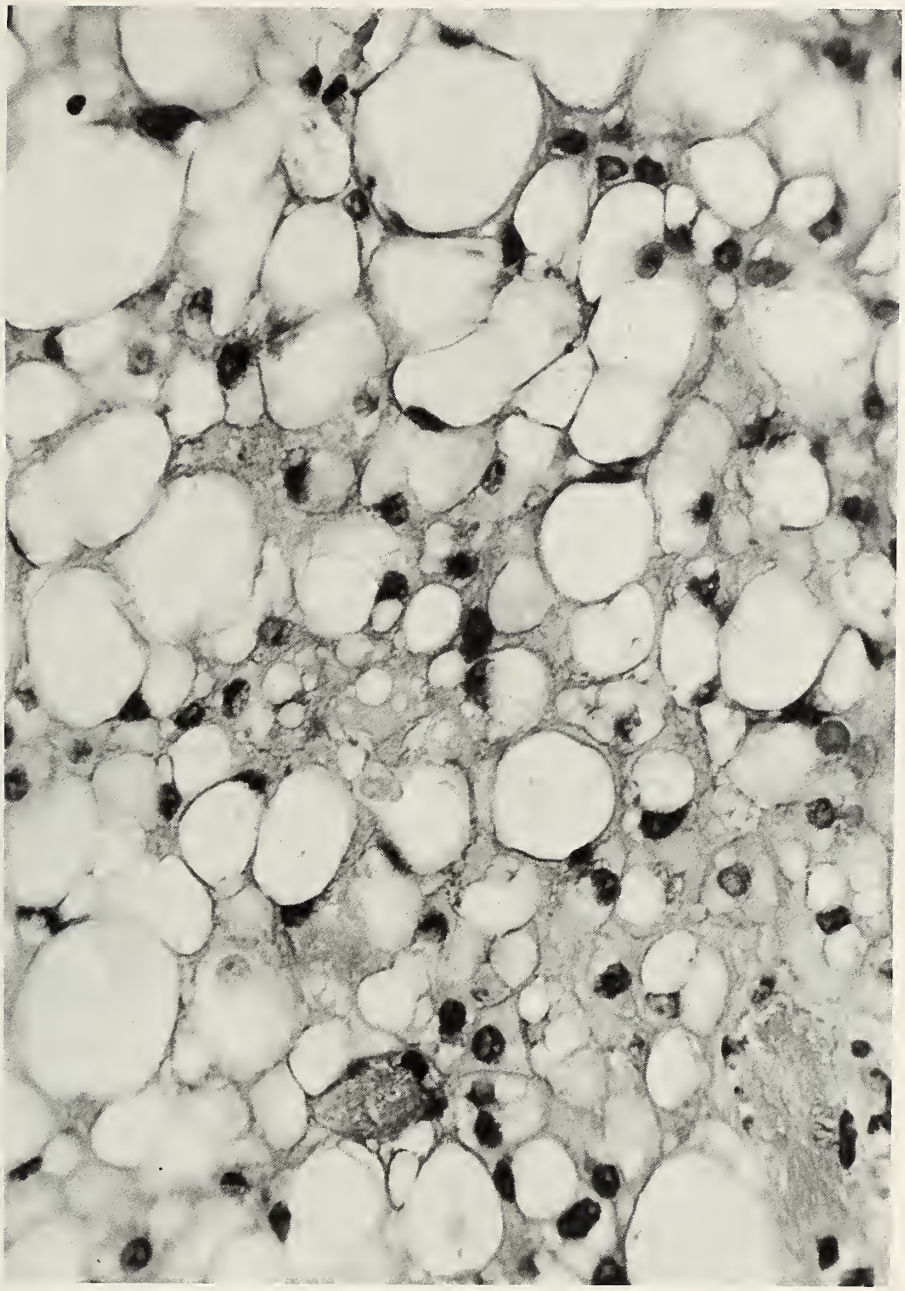


Figure 60. Note the large vacuolated cells. Nuclei are uniform, and no mitoses are seen. × 600. A. F. I. P. Acc. No. 219023–71.

CHORDOMA

(Figures 61–64 are from the same case)

Figure 61. A roentgenogram, taken in 1941, showing the pelvis in a 50-year-old patient, who had had rectal pain and progressive constipation for 18 months. He developed acute urinary retention. The roentgenogram demonstrates a retention catheter in place, with evidence of prominent bone destruction of the sacrum. This patient received only irradiation therapy, 2,000 r to each of four large fields. A. F. I. P. Acc. No. 219023–72.

Figure 62. This roentgenogram taken in 1948 demonstrates some recalcification of the sacrum, but a large bony defect is still present. On rectal examination in 1952 a small nodule of apparently persistent tumor could still be felt. A. F. I. P. Acc. No. 219023–73.

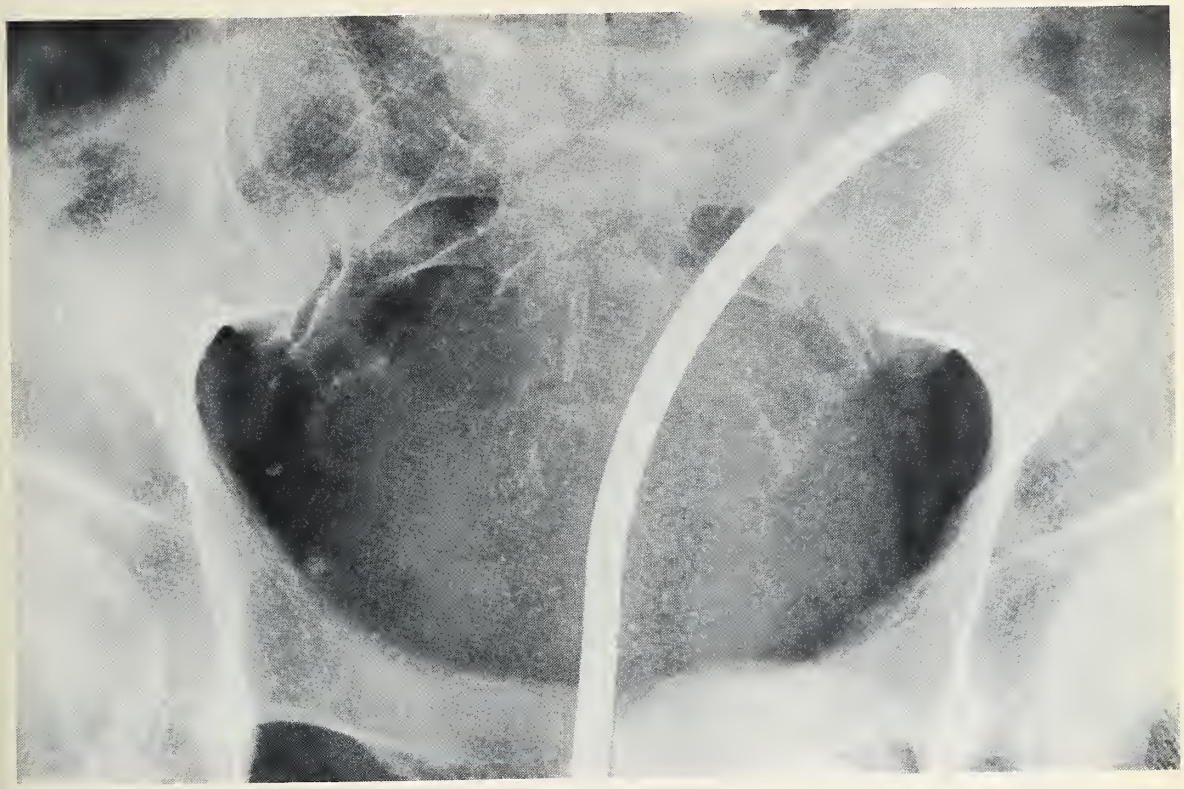


Fig. 61

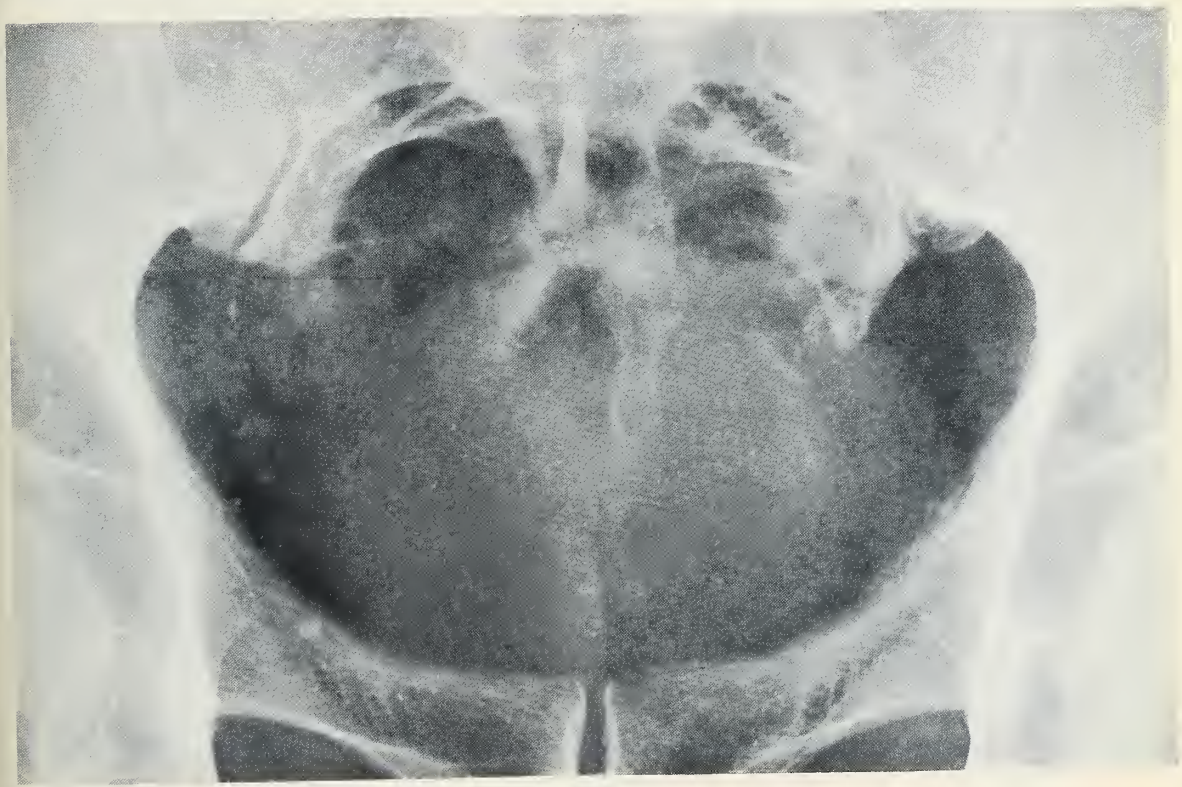


Fig. 62

CHORDOMA

(Figures 61–64 are from the same case)

Figures 63 and 64. Photomicrographs of the typical cells of the chordoma shown by preceding roentgenograms. × 1200 and × 2000. A. F. I. P. Acc. Nos. 219023–48 and 219023–49.

Fig. 63

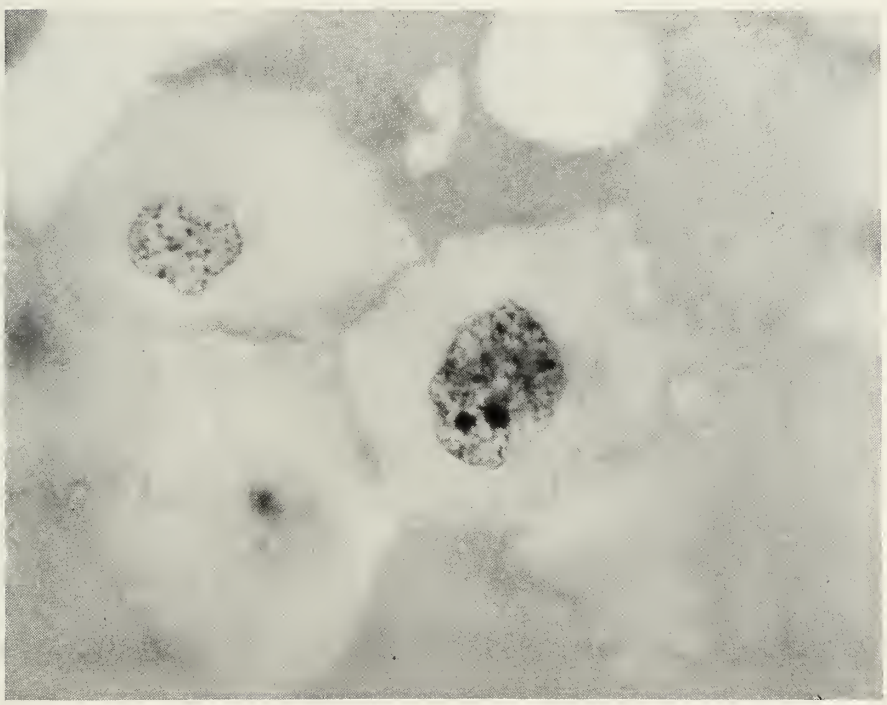


Fig. 64

little or no vacuolation, it is considered by Faust, Gilmore, and Mudgett to be more malignant than the highly vacuolated tumor.

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METASTATIC TUMORS

Metastatic malignant tumors involving the retroperitoneal area are a common clinical and pathologic finding, and are even more common as a post-mortem finding as part of a generalized process. The clinical symptoms which suggest a primary retroperitoneal tumor may represent a metastatic process. The malignant testicular tumors invade this area, and usually involve the nodes along the aorta (pl. IV–C). Metastatic carcinoma of the cervix may form a mass in the midabdomen. Hodgkin's disease, lymphosarcoma, and carcinoma of the stomach and pancreas often produce massive involvement of the aortic nodes. Chondrosarcoma of the bones of the pelvis may directly invade the retroperitoneal space.

TUMORS OF THE MESENTERY AND OMENTUM

(Originally planned as Fascicle 23)

EMBRYOLOGIC DEVELOPMENT OF THE MESENTERY

The embryology of the mesentery and its morphologic relationship to neighboring structures is of unquestioned significance in the production of certain types, and probably of the majority, of these tumors. The primitive alimentary canal is developed within the cavity of the coelom, within the median partition which separates the right coelomic space from the left. This median partition suspends the primitive gut from the dorsal body wall and forms the dorsal mesentery, whereas that part of the partition which fixes the digestive tract to the ventral body wall constitutes the primitive ventral mesentery.

From an embryologic standpoint, it is conceivable that remnants of the wolffian or müllerian ducts or segregated remnants of the genital gland can become lodged within the mesentery. The genital ridge develops lateral to the mesentery of the gut. The genital and the wolffian bodies each have their own mesenteries, but these mesenteries have a common attachment—the common urogenital mesentery. The duct of the wolffian body is situated in the lateral margin of the genital ridge, dorsal to the müllerian duct. It is possible that portions of these embryonic structures might migrate in the retroperitoneal tissues to invade the mesentery of the bowel, and act as nuclei of mesenteric tumors in postnatal life.

BENIGN TUMORS

Cystic tumors of the mesentery are more common than the solid type (Rankin and Major). In the benign tumor group cystic **lymphangiomas** are most common (figs. 65, 66). These tumors do not become malignant. They may involve a wide area of the mesentery. Grossly they form numerous cystic spaces of variable size. Dowd believed that mesenteric cysts might have something to do with sequestration from embryonic genital glands which could be included within the folds of the mesentery, mesocolon, or mesorectum. **Hemorrhagic** cysts of the mesentery are probably traumatic (Moynihan; figs. 67, 68). In Courty and Falala's review of 216 solid tumors of the mesentery, the commonest benign tumor was a **lipoma. Hemangiomas,** vascular **leiomyomas** (Hartz and van de Stadt), and even **ganglioneuromas** (Bland-Sutton) have been reported. (See table III.) We have seen a desmoid tumor involving the mesentery following trauma.

LYMPHANGIOMA OF OMENTUM

(Figures 65 and 66 are from the same case*)

Figure 65. External surface of a large multiloculated cystic lymphangioma of the omentum. A. F. I. P. Acc. No. 219023–78.

Figure 66. Cut surface of the lymphangioma. A. F. I. P. Acc. No. 219023–79.

*Courtesy of Dr. D. H. Kaump, Providence Hospital, Detroit, Mich.

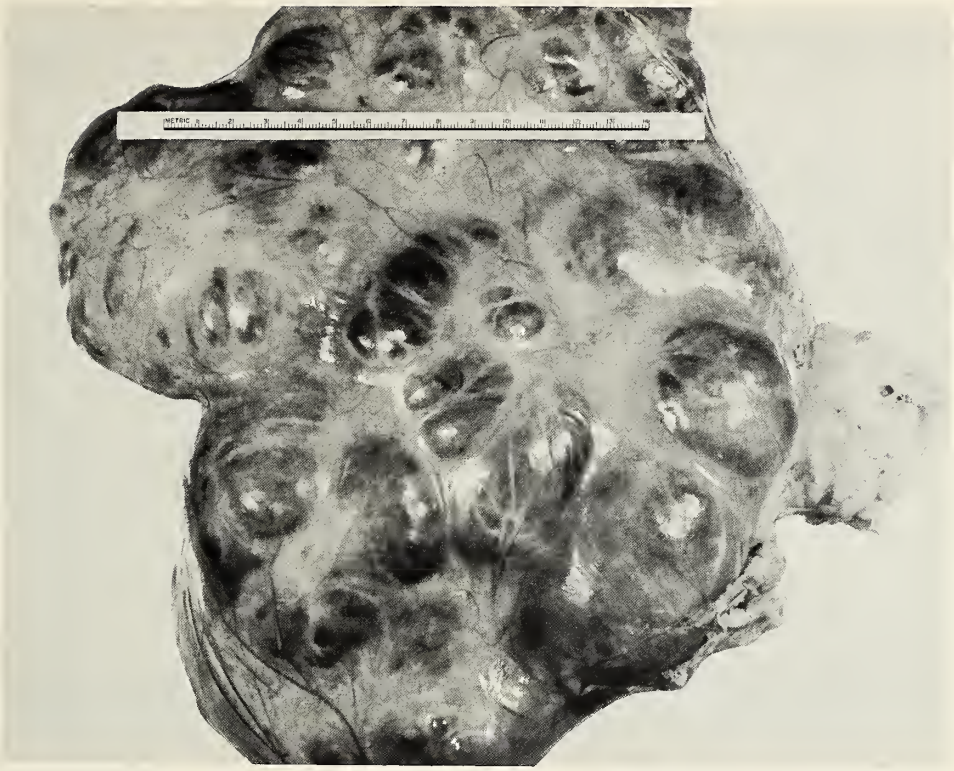

Fig. 65

Fig. 66

HEMORRHAGIC CYST

(Figures 67 and 68 are from the same case)

Figure 67. External surface of a large hemorrhagic cyst of the mesentery. A. F. I. P. Acc. No. 219023–76.

Figure 68. Cut surface of the cyst. A. F. I. P. Acc. No. 219023–77.

Fig. 67

Fig. 68

FIBROSARCOMA OF MESENTERY

(Figures 69 and 70 are from the same case*)

Figure 69. Fibrosarcoma of the mesentery of the jejunum. This was an infiltrating growth, 7.5 cm. in diameter, that had deformed the intestinal wall, to which it was closely adherent. There was no recurrence of symptoms seven years after resection. A. F. I. P. Acc. No. 219023–74.

Figure 70. Photomicrograph showing the well differentiated fibroblastic cells. × 470. A. F. I. P. Acc. No. 219023–75.

*From Stout, A. P. Fibrosarcoma. The malignant tumor of fibroblasts. Cancer, 1: 30–63, 1948. Figures 13 and 14 are our figures 69 and 70.

Fig. 69

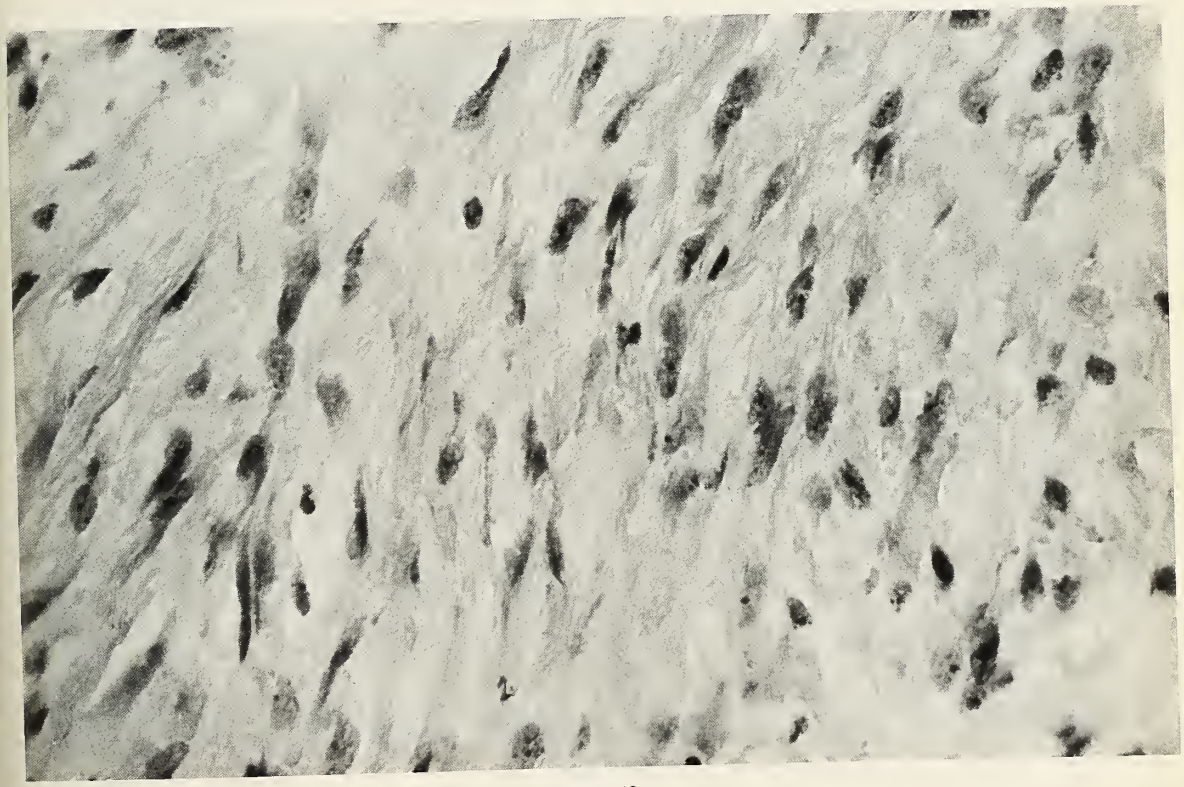

Fig. 70

Table III

BENIGN TUMORS ARISING IN THE MESENTERY OR OMENTUM
COLUMBIA UNIVERSITY, 1905 TO 1951 INCLUSIVE*

	Mesenteric	Omental
Lipoma	2	5
Leiomyoma	1	4
Lymphangioma	3	.....
Neurofibroma	1	.....
Total	7	9

*Courtesy Arthur Purdy Stout, M. D.

MALIGNANT TUMORS

The most common malignant tumors of the omentum are the **metastatic tumors.** Such tumors frequently arise from the stomach, the pancreas, and the ovaries, and such metastases may also appear from many other organs. The primary malignant tumors of the mesentery and omentum are more frequently **liposarcomas** than any others (pl. IV–B). Stout has reported six well differentiated **fibrosarcomas** of the mesentery (figs. 69, 70). None of his six cases had metastasized at the time of the report (see table IV).

Table IV

MALIGNANT TUMORS ARISING IN THE MESENTERY OR OMENTUM
COLUMBIA UNIVERSITY, 1905 TO 1951 INCLUSIVE*

	Mesenteric	Omental
Fibrosarcoma	6	.....
Liposarcoma	2	**2
Leiomyosarcoma	2	4
Lymphosarcoma	2	2
Hemangiopericytoma	1	2
Plasmocytoma	1	.....
Undiagnosed tumors	1	4
Total	15	14

*Courtesy Arthur Purdy Stout, M. D.
**3 more attached to kidney capsule

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PLATE V

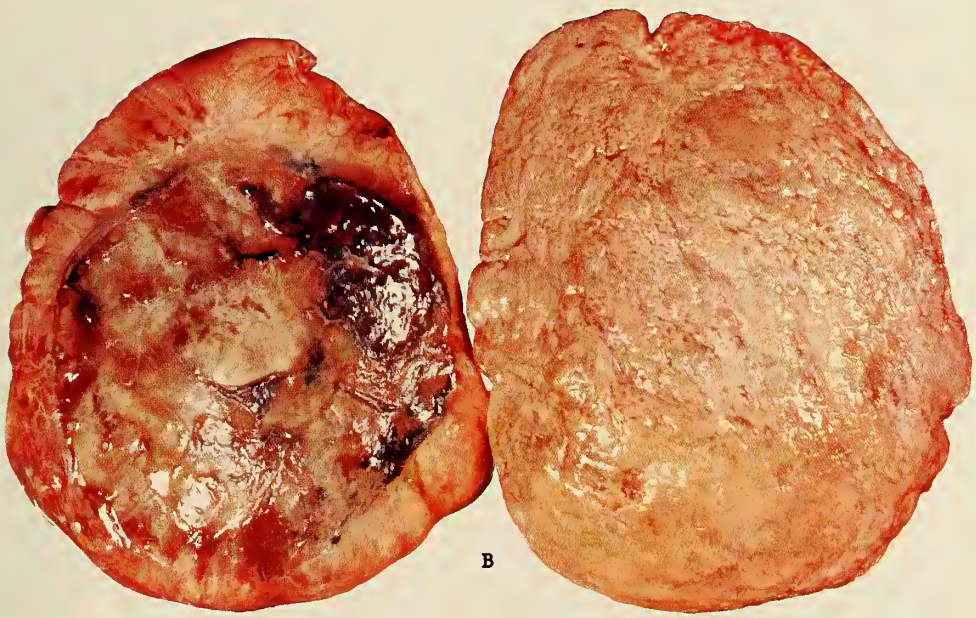
MESOTHELIOMA

(Plate V–A and figure 75 are from the same case)

A. Fibrous type of mesothelioma of peritoneum. Same case as figure 75. (Contributed by Dr. A. P. Stout, Columbia University, College of Physicians and Surgeons, New York, N. Y.) A. F. I. P. Acc. No. 219023–C1.

B. Fibrous type of mesothelioma of the pleura. Note the striking resemblance between this tumor and that in plate V–A. Both are solid, grayish pink in color, without evidence of necrosis. A. F. I. P. Acc. No. 219023–24.

A



B

PLATE V

TUMORS OF THE PERITONEUM
(Originally planned as Fascicle 23)

Introduction

The primary peritoneal neoplasms have an extremely variable microscopic picture because of their embryonic background. It must be remembered that from condensations of the mesothelium in the embryo, the mesonephros and metanephros, wolffian body, genital ridge, and all the dependent urogenital structures are derived (Keasbey, 1947). Furthermore, the coelomic cavity is lined by ciliated mesothelium when the development of the excretory apparatus is rudimentary. Even squamous metaplasia of the peritoneum can occur with a formation of cystic nodules (Crome), and we also have seen such cases. There is definite metaplasia to the squamous type with the formation of intercellular bridges (fig. 71). It would, therefore, be theoretically possible to have even a primary squamous carcinoma of the peritoneum. In this regard, it is also of interest to mention the theory that endometriosis may arise from metaplasia. The lining mucous membrane of all parts of the müllerian canal (tubes, uterus, and vagina), as well as the germinal epithelium of the ovary and pelvic peritoneum, comes from the coelomic cavity (Novak).

Primary tumors of the mesothelial lining cells of the peritoneal cavity are rare. Unfortunately in the past such tumors were poorly described and often were metastatic rather than primary neoplasms of the peritoneum. There is no doubt of the occurrence of mesothelioma of the pleura (Liebow*); and occasionally we see synovial sarcomas (Haagensen and Stout), and even tumors arising from the pericardium.

The mesothelioma originating from the tunica vaginalis testis has been described in Fascicle 32, "Tumors of the Male Sex Organs."** The tunica vaginalis testis is derived from a downgrowth of the coelomic cavity and is in reality a part of the peritoneal cavity. However, as far as can be determined from a search of the literature, the tumors of the mesothelium of this area, although they have considerable variation in their pattern, are invariably benign (Masson, Riopelle, and Simard; Golden and Ash; Evans). The tumors of the peritoneal surface of the fallopian tube (Evans), the counterpart of these tumors, can arise as locally diffuse growths from the peritoneal surface of the uterus (Sternberg). This discussion, however, will be confined to the primary

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* Fascicle 17, "Tumors of the Lower Respiratory Tract"

**Section on Adenomatoid Tumors, p. 127.

SQUAMOUS METAPLASIA

Figure 71. Squamous metaplasia of mesothelium.

A. A squamous metaplasia of the peritoneum. Intercellular bridges are present. × 360. A. F. I. P. Acc. No. 219023–100.

B.* Squamous metaplasia of the pleura. The presence of the granular layer and keratinization are unusual. × 200. A. F. I. P. Acc. No. 583958–1.

C.* Higher power of the squamous metaplasia illustrated in figure 71–B. Intercellular bridges are clearly depicted. × 1,000. A. F. I. P. Acc. No. 583958–2.

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*B and C are from the same case. Called to my attention by Dr. Franz Liedler, chief pathologist, Veterans Hospital, Jefferson Barracks, Mo. Material on this case was subsequently sent to the Armed Forces Institute of Pathology.

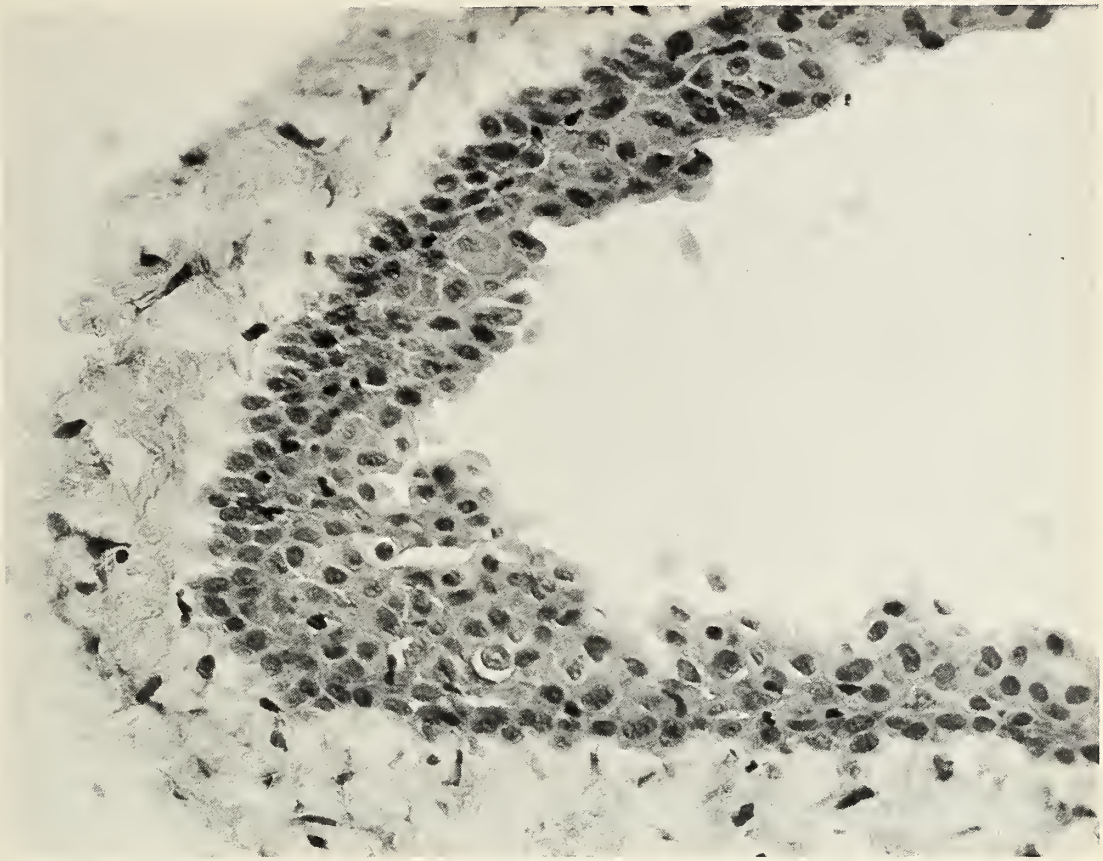


Fig. 71 A

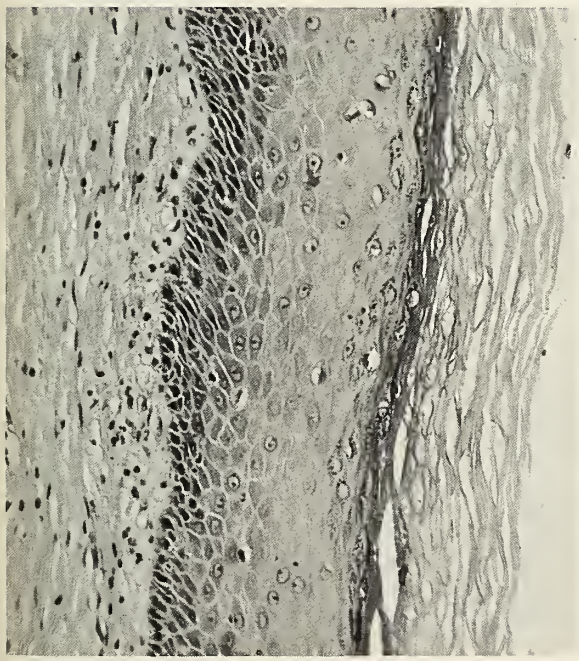


Fig. 71 B

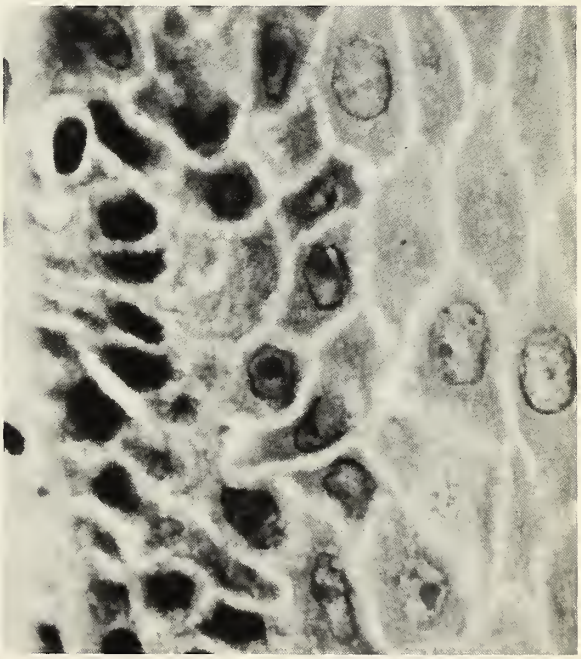


Fig. 71 C

tumors within the peritoneal cavity. Mesotheliomas can also occur in horses and cows, and these neoplasms are similar to those in man (Jackson; figs. 72–74).

The cells of the peritoneum, like those of the synovium, secrete a substance called hyaluronic acid. This substance has been isolated from viscous pleural fluid of a patient with a malignant tumor of the pleura and peritoneum. In this case, Meyer and Chaffee spoke of the impossibility of staining any mucin within cells by the use of the mucicarmin stain. There were vacuoles present which were shown to represent fat globules since they were stained by Scharlach R. In this same patient, hyaluronidase caused liquefaction of the viscid fluid. We have not been able to demonstrate epithelial mucin in primary mesothelial tumors. In metastatic tumors involving the peritoneum and simulating a primary tumor, the mucin stain is often positive. This occurs particularly with metastatic adenocarcinoma from the pancreas, gall bladder, and stomach.

Classification

The following classification is made mainly for the purpose of organizing the discussion, since transition from one type to another may occur. The distinction between a benign and a malignant tumor may be difficult. Stout's recorded experience and classification are summarized in table V.

Classification of tumors of the peritoneum:

- Benign
  - Fibrous, localized or diffuse
  - Papillary, localized or diffuse
- Malignant
  - Papillary, localized or diffuse
  - Tubular (resembling synovial sarcoma), localized or diffuse

The distribution of these types varies in the pleura and peritoneum. Stout's series is the largest one recorded and the distribution of the cases is in table V.

Mesothelioma

The fibrous type of mesothelioma is an extremely rare neoplasm with a distinctive gross appearance (Stout; pl. V–A, B). In the pleural cavity these neoplasms have been considered by some to be the end results of a low grade inflammatory process with organization of the exudate (Brown). The lack of antecedent history of inflammation and the presence of similar tumors within the peritoneal cavity would negate this concept. The tumor is firm, encapsulated, solid, and on section has a grayish pink surface without necrosis or cystic degeneration. Counterparts of these neoplasms have been described in the pleural cavity (Stout, 1951) and the best proof of their origin is in the tissue culture studies of Murray. Murray's work was later confirmed by Sano, Weiss, and Gault.

Table V*

MESOTHELIOMA OF THE PLEURA, PERITONEUM, AND PERICARDIUM
LABORATORY OF SURGICAL PATHOLOGY, COLUMBIA UNIVERSITY,
JULY 1919 TO DECEMBER 1951

	Pleura	Peritoneum	Pericardium	
Benign Mesotheliomas:				
Solitary				
Fibrous	35	3	.....	38
Tubular	3	**13	.....	16
Diffuse				
Fibrous	2	.....	.....	2
Tubular	....	.....	.....	...
Malignant Mesotheliomas:				
Solitary				
Fibrous	17	5	.....	22
Tubular	1	8	.....	9
Diffuse				
Fibrous	....	1	2	3
Tubular	***4	***21	2	27
Total	62	51	4	117

*Stout, A. P. Mesothelioma of the Pleura and Peritoneum (to be published). This table includes cases received from 64 hospitals in 21 states.
**Epididymis 6, uterus 3, fallopian tube 3, sigmoid colon 1.
***One additional case involving both pleura and peritoneum occurred in an old cow.

The microscopic pattern of the fibrous type reveals adult fibrous tissue forming a disordered tangle of fibers with abundant reticulin (fig. 75). Wide bands of hyalinization may be seen. No tubular areas are present. We have not seen a good example of a diffuse benign fibrous mesothelioma. Local excision of this circumscribed tumor, whether it arises within the pleura or peritoneum, is curative.

Papillary tumors of the peritoneum are the commonest form of primary peritoneal tumor, and are often mimicked by metastatic papillary serous carcinomas of the ovary (figs. 89, 90). However, this type of neoplasm also occurs in the male. In the benign form, small papillary masses (fig. 76) form on the surface of the peritoneum. These masses are most commonly located on the pelvic peritoneum, diaphragm, mesentery, or on the surface of the intestine. In its earliest form this tumor causes nodulation of the peritoneum with the formation of papillary masses which may become conglomerate. These lesions may occupy a small or large area of the peritoneal surface, and

MESOTHELIOMA OF COW

Figure 72.* Note the similarity in gross appearance to figure 77. A. F. I. P. Acc. No. 62734.

Figure 73.* This is a diffuse lesion involving the peritoneum. A. F. I. P. Acc. No. 62733.

*From the Armed Forces Institute of Pathology.

Fig. 72

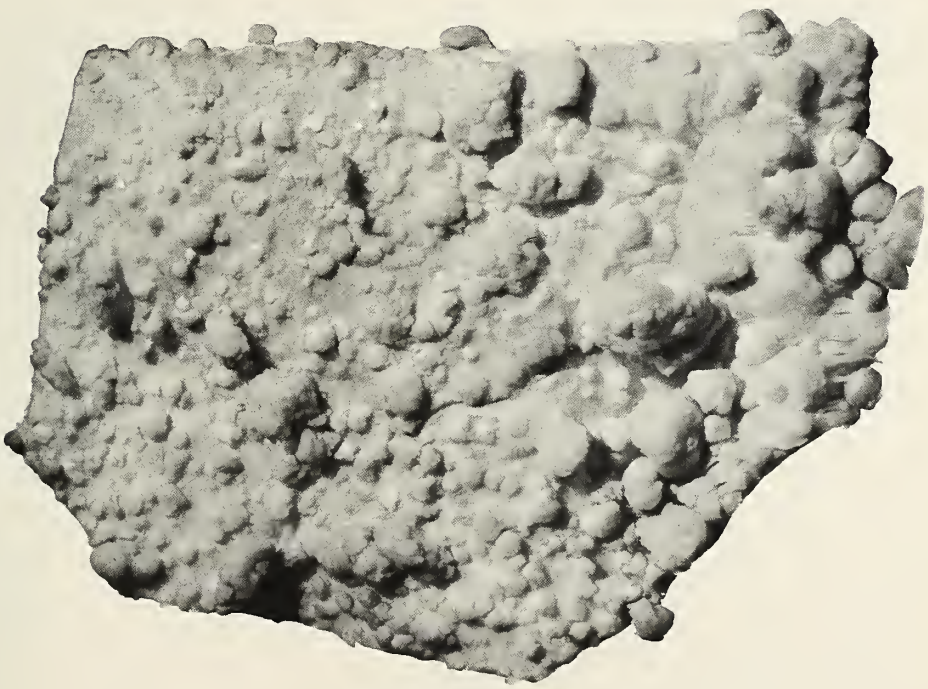


Fig. 73

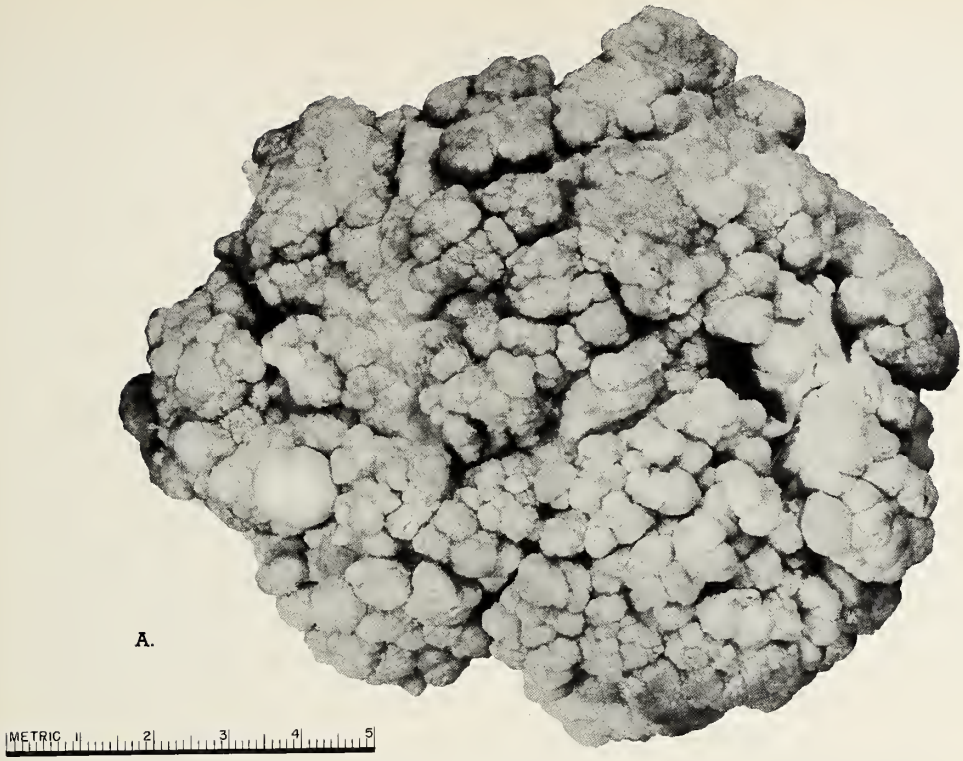
MESOTHELIOMA OF COW

Figure 74.* Malignant papillary mesothelioma.

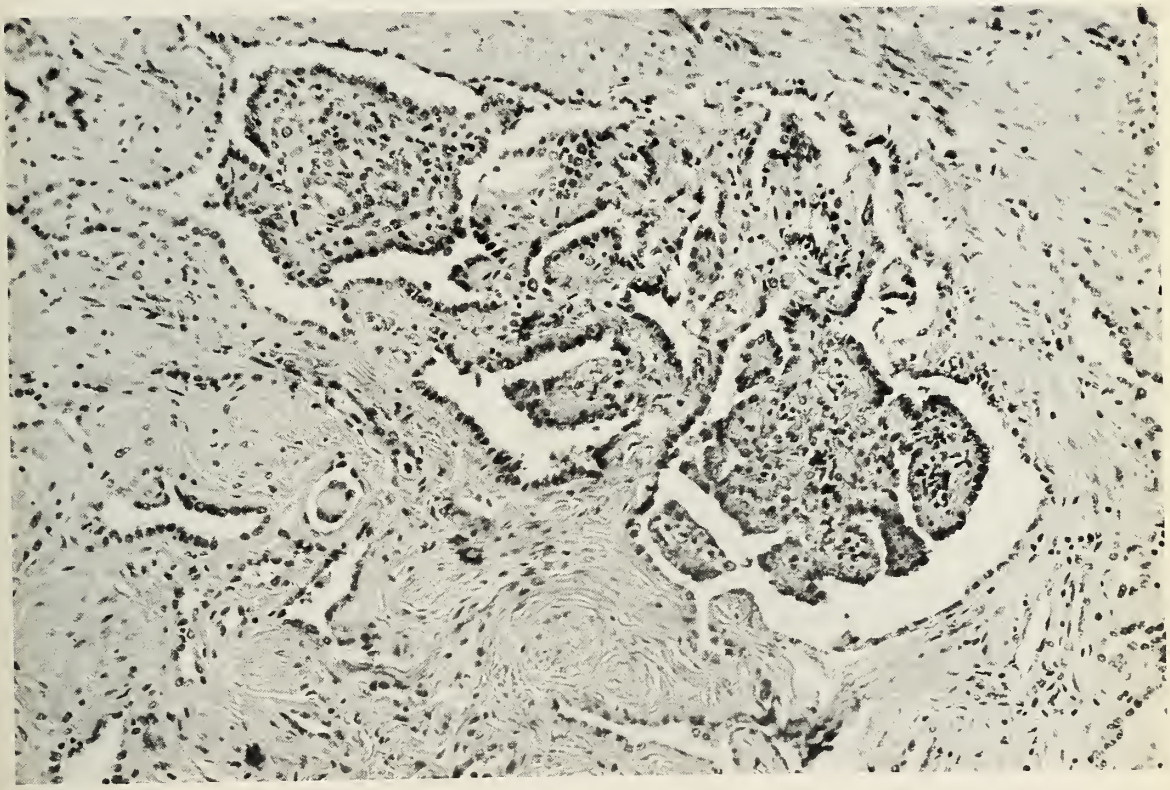
A. Gross photograph of a papillary mesothelioma of the mesentery of a 7-year-old cow. Similar tumor was present in the pleura, and the bronchial lymph nodes were also involved. A. F. I. P. Acc. No. 216270–1.

B. Low power photomicrograph of the tumor illustrated in figure 74–A. × 200. A. F. I. P. Acc. No. 216270–2.

*Case contributed to the Armed Forces Institute of Pathology by Dr. C. L. Davis, Denver, Colo.



A.



B.

Fig. 74

MESOTHELIOMA

(Figure 75 and Plate V–A are from the same case)

Figure 75. Photomicrograph of the fibrous type mesothelioma demonstrated in plate V–A. × 650. (Courtesy of Dr. A. P. Stout, Columbia University, College of Physicians and Surgeons, New York, N. Y.) A. F. I. P. Acc. No. 219023–101.

Figure 76. Photomicrograph of a benign papillary mesothelioma. Note typical lining cells with loose vascular stroma. Lesion found incidentally at operation. × 120. A. F. I. P. Acc. No. 512591.

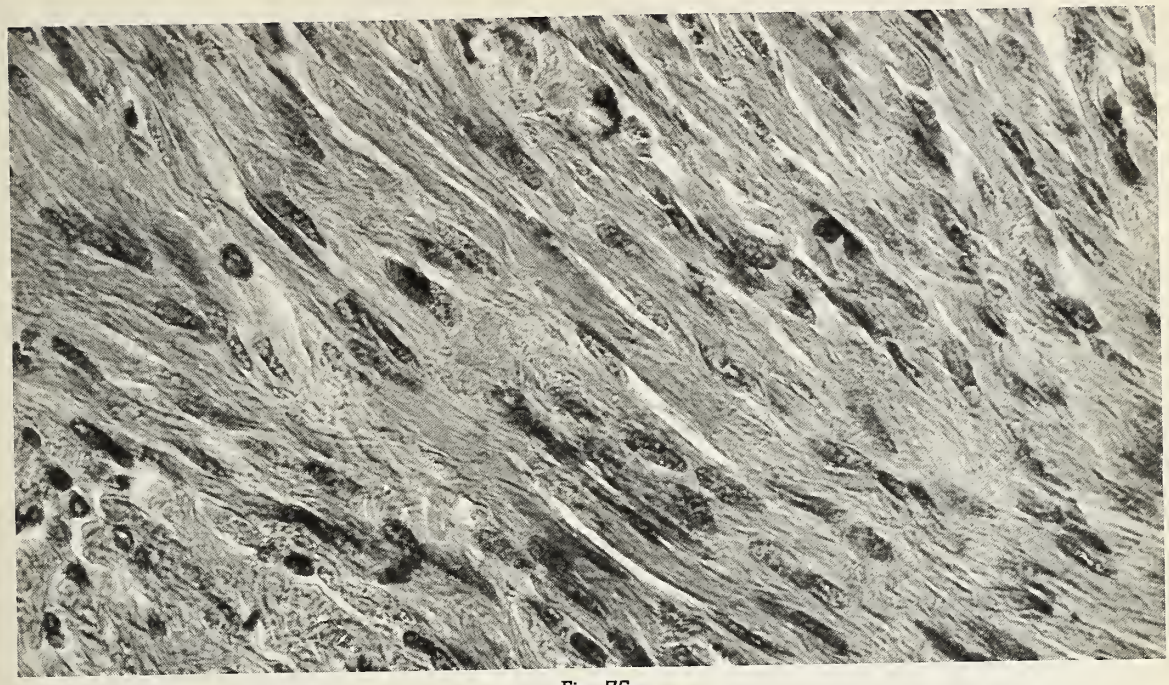


Fig. 75

Fig. 76

it is probable that implantation as well as multiple foci of origin play a role in their development. Cases of this nature have been described as incidental findings at autopsy. Wells reported the case of a 78-year-old man who died following prostatectomy. At autopsy there were multiple small papillary tumors scattered on the surface of the peritoneum, diaphragm, and mesentery. Keasbey (unpublished data) observed a similar case with involvement of the peritoneal cavity and the tunica vaginalis testis in a man 69 years of age who died following a traumatic cerebral hemorrhage. These papillary tumors or distinct projections are associated at times with considerable increase of subserosal connective tissue. Rarely such an involvement may include not only the peritoneum but also the pleura. A case reported by Yoshida occurred in a 45-year-old man who died of unrelated causes. There was considerable viscid fluid within the peritoneal cavity and there were no metastases.

We also have seen a similar case of long duration in a man, age 44 (Skilling). This patient over a 4-year period had repeated accumulation of fluid in both the peritoneal and the pleural cavities; and this fluid contained large masses of mesothelial cells that were not interpreted as tumor because these cells were extremely well differentiated (fig. 81). The fluid found in this case was rather viscid. The pleural cavity was obliterated and thickened (fig. 78). The peritoneal cavity was almost completely obliterated by dense fibrous tissue (fig. 77). The parietal and visceral peritoneum were thickened and nodular. Microscopically these nodules were made up of papillary projections with a central fibrous tissue core (figs. 79, 80). The papillary projections showed some branching into primary or secondary branches and were lined by cuboidal or columnar cells with pink-staining cytoplasm and well defined nuclei. Individual cells were well differentiated, nucleoli were prominent, and mitotic figures were rare (fig. 82). This appearance of the cells explains the impossibility of making this diagnosis in the examination of ascitic or pleural fluid (fig. 81). There were no distant metastases, but there was involvement of some regional lymph nodes by direct extension. In another case* there was extensive peritoneal but no pleural involvement. However, there was extension of the tumor into the wall of the intestine, stomach (figs. 83, 84), diaphragm, and hilus of the spleen.

The clinical course and the gross and microscopic findings are typical of the papillary type of mesothelioma. It is probable that the clinical history is even longer than is suggested here. It is also possible that some cases which cause the death of the patient may begin as an apparently localized benign process. The benign forms of this process are only rarely seen except as incidental findings either at operation or at autopsy. It is possible that some

*Called to my attention by Dr. Franz Leidler, Chief pathologist, Veteran's Hospital, Jefferson Barracks, Mo. Material on this case was subsequently sent to the Armed Forces Institute of Pathology.

remain benign, for Keasbey (unpublished data) saw an early case in a woman, age 27, in whom the lesion was found incidentally at operation and who was free of disease 11 years later.

Such tumors can at first be localized, and if present in a critical area may produce symptoms. In a 73-year-old man a localized mesothelioma produced enough involvement to cause intestinal obstruction. This tumor was not recognized grossly as a mesothelioma, and 85 cm. of ileum was resected. The lesion was a papillary mesothelioma that had involved the wall of the bowel but that did not involve the mucosa (figs. 85, 86). Several years later this patient died with evidence of diffuse peritoneal disease but without distant metastases. When a malignant papillary tumor first appears with only a few nodules, it is usually thought at operation to be tuberculosis.

In the terminal evolution of these papillary tumors the peritoneal cavity is finally obliterated, and no more fluid is secreted. All the organs are matted together and have to be removed en masse.

Finally we have seen a young man, age 24, who had a localized tumor of the peritoneum. It was resected and had all the microscopic characteristics of synovial sarcoma. There were pseudoglandular spaces and a sarcomatous stroma (figs. 87, 88). This patient later died and was autopsied by Dr. Frank McKee of the Genesee Hospital, Rochester, New York. In view of the capacity of the mesothelium of the peritoneal cavity to undergo diverse change, it is possible that highly undifferentiated malignant neoplasms occur there. However, in view of our present uncertainty we do not feel justified in illustrating such doubtful cases until better evidence of origin is available.

Metastatic Carcinoma

Metastatic cancer of the peritoneum like metastatic cancer of the pleura can imitate with great exactitude, both grossly and microscopically, primary tumors of the peritoneum. In fact, some eminent pathologists still hold that primary peritoneal tumors do not exist (Willis). Ovarian neoplasms, particularly the serous type, can imitate the papillary type of primary peritoneal tumor (figs. 89, 90). Imitation of the diffuse type of mesothelioma occurs commonly. Usually a cursory review of the pertinent facts of the clinical and physical findings will reveal some obvious source of a primary tumor. Careful postmortem examination may also reveal the primary. The most common primary tumors are from the ovary, pancreas, stomach, large bowel, and lung. It can be argued by those who believe that this entity does not exist that if in a diffuse peritoneal tumor a primary malignant neoplasm from some other source is not demonstrated, this means only that the search was not thorough enough. There are, however, curable primary benign peritoneal tumors simulating the malignant types (Stout, 1950 and 1951). Tissue culture studies also have produced evidence in support of a mesothelial origin (Stout and Murray;

MESOTHELIOMA

(Figures 77–82 are from the same case)

Figure 77. Photograph of gross specimen of a diffuse type of malignant mesothelioma. Note extreme fibrosis with shortening of the mesentery. A. F. I. P. Acc. No. 219023–80.

Figure 78. Gross specimen of lung. Note extreme fibrous thickening of the pleura by tumor. A. F. I. P. Acc. No. 219023–81.

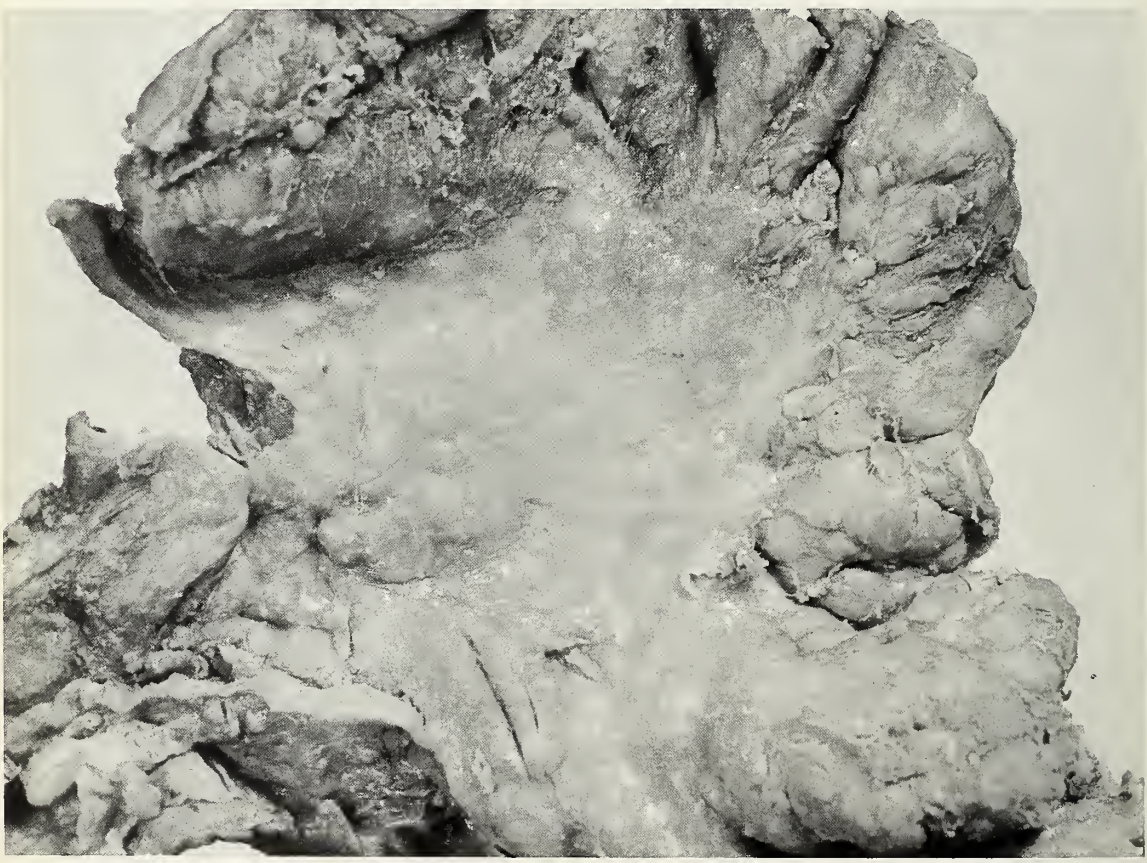


Fig. 77

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MESOTHELIOMA

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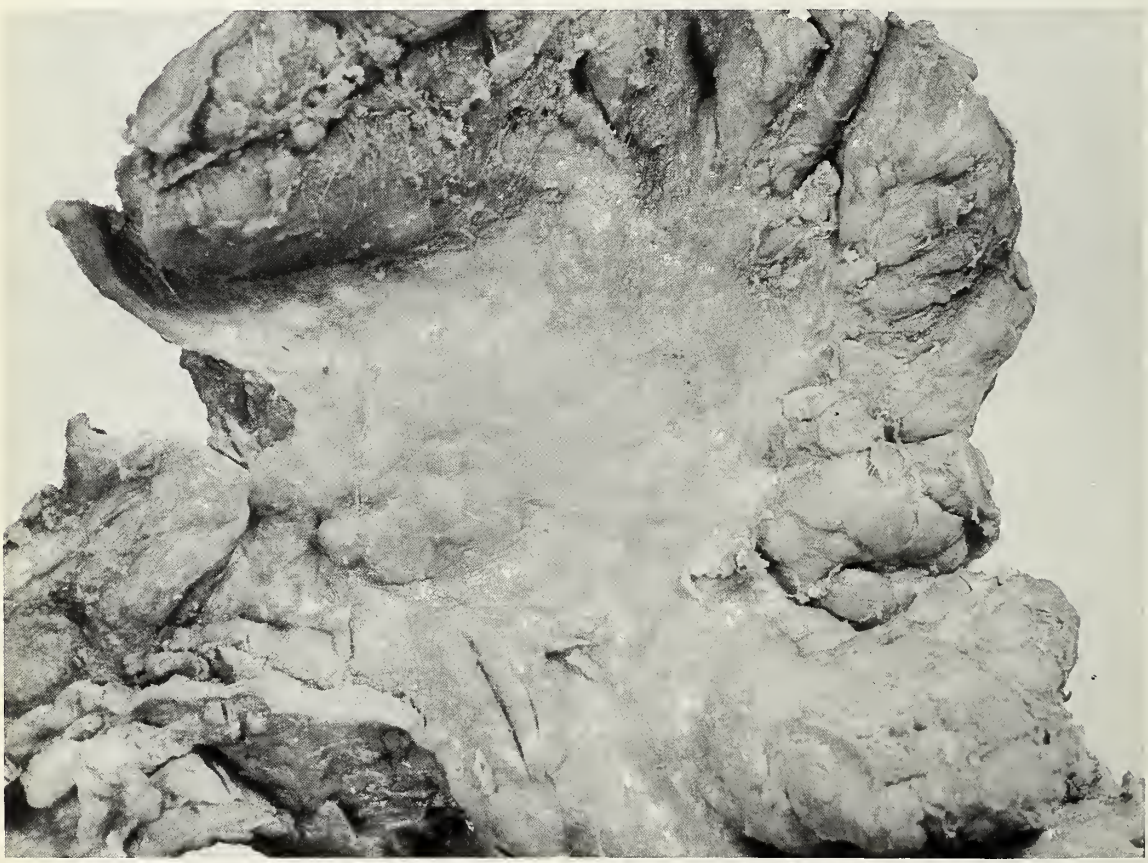

Fig. 77

Fig. 78

MESOTHELIOMA

(Figures 77–82 are from the same case)

Figure 79. Photomicrograph to demonstrate the papillary pattern supported by cores of connective tissue. × 360. A. F. I. P. Acc. No. 219023–82.

Figure 80. Higher magnification to show detail of the malignant mesothelioma. Note irregularity of cells with rather prominent nuclei. × 560. A. F. I. P. Acc. No. 219023–83.

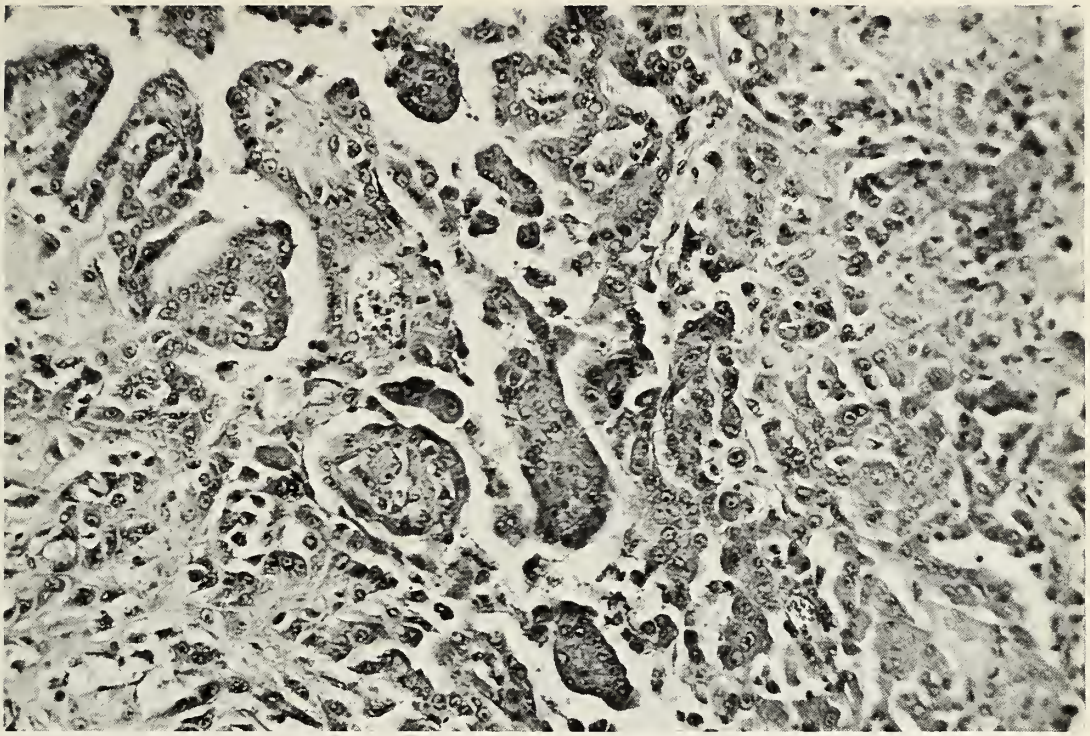

Fig. 79

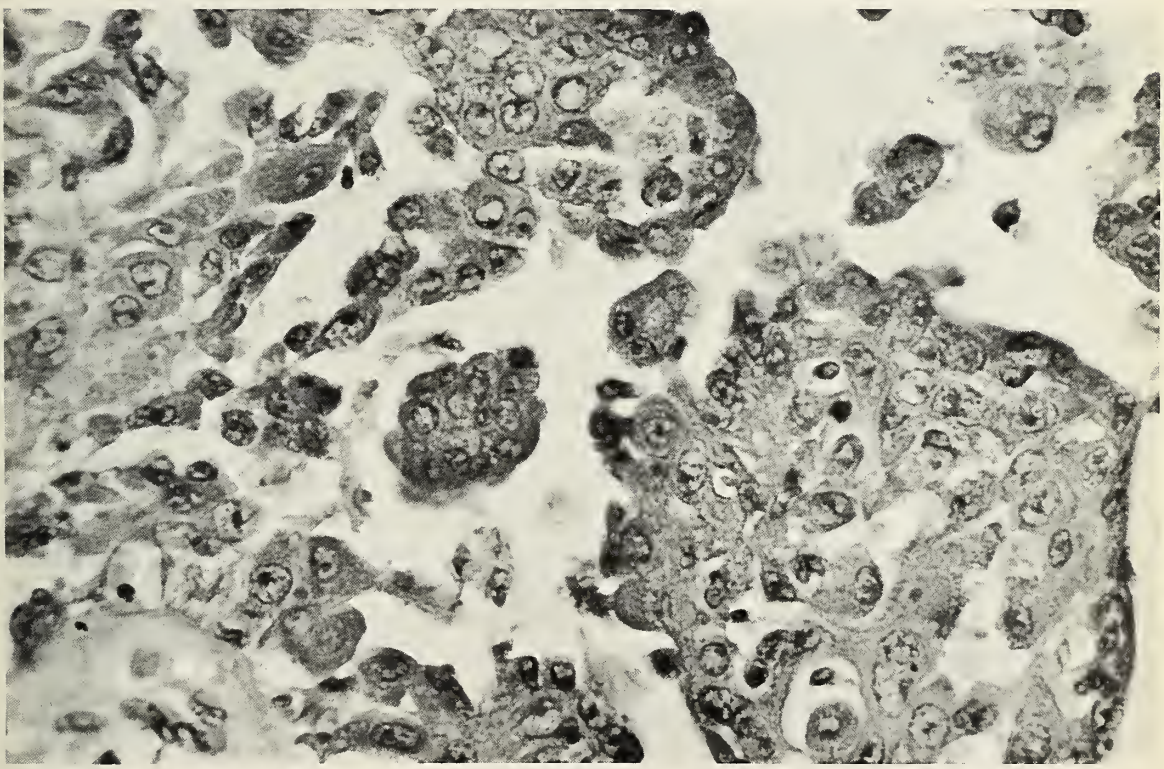

Fig. 80

MESOTHELIOMA

(Figures 77–82 are from the same case)

Figure 81. Ascitic fluid sediment of well differentiated cells from a case of papillary mesothelioma. Note prominent nuclei. × 900. A. F. I. P. Acc. No. 219023–84.

Figure 82. Photomicrograph of a histologic section of the malignant mesothelioma to show abundant cytoplasm, prominent nuclei, and rather prominent nucleoli. Compare with figure 81. × 800. A. F. I. P. Acc. No. 219023–13.

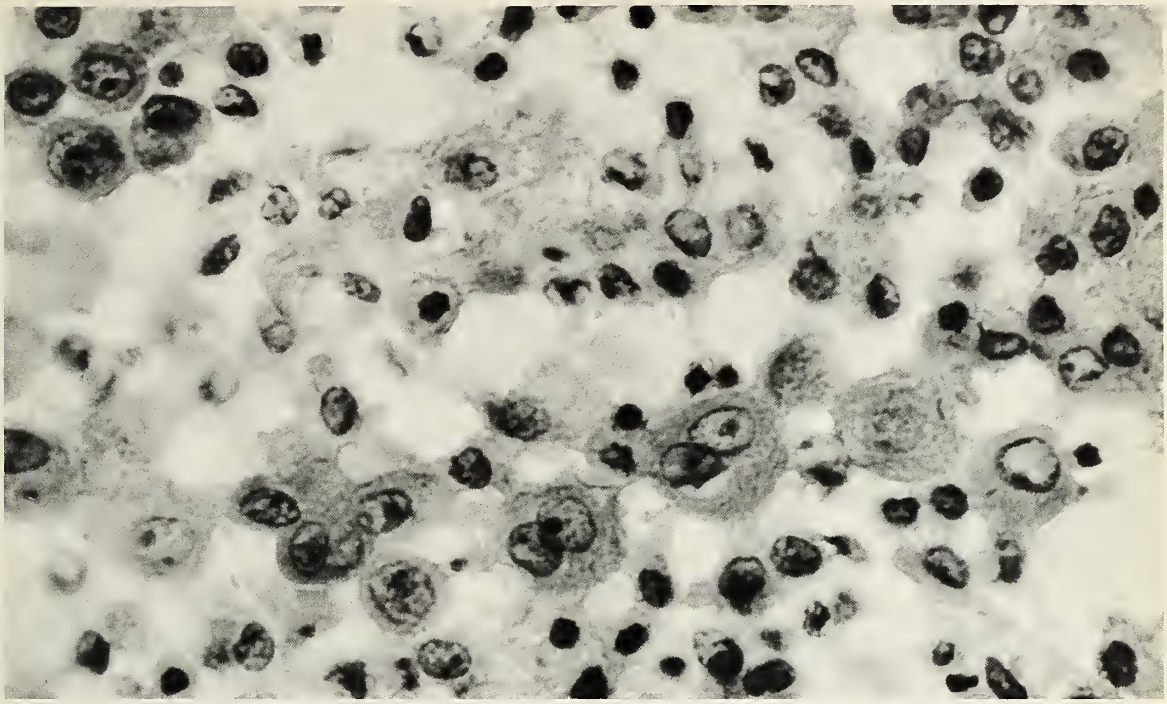

Fig. 81

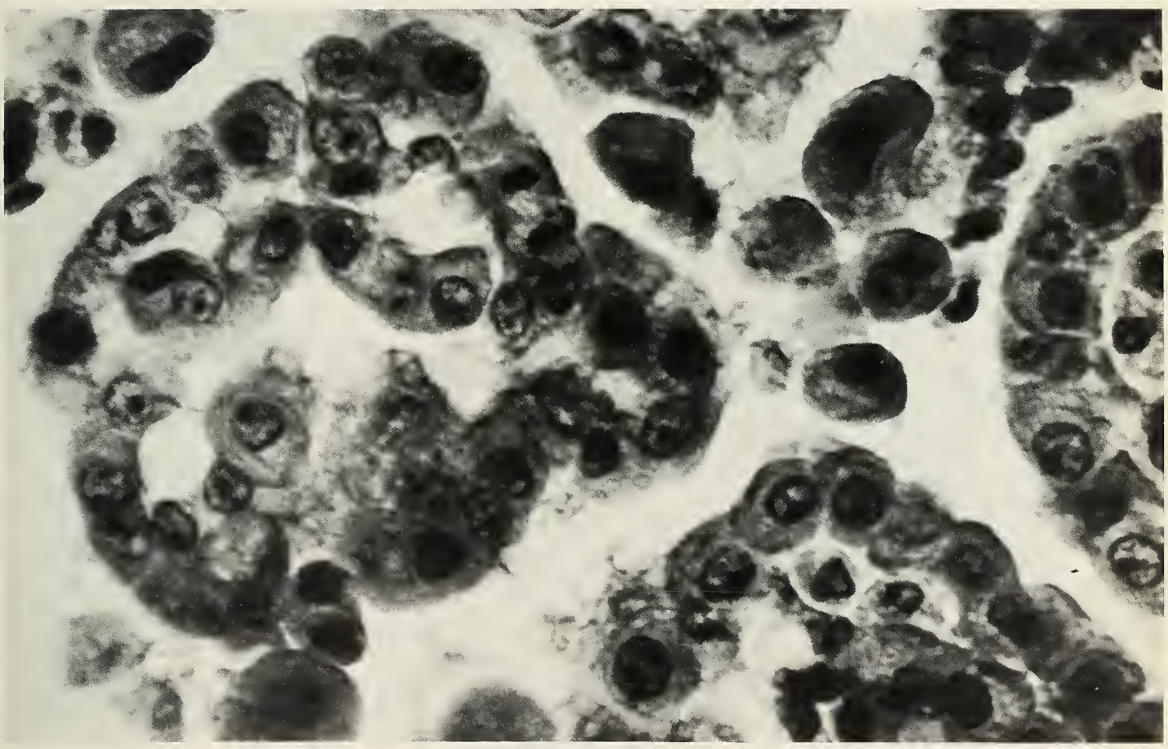

Fig. 82

MESOTHELIOMA

(Figures 83 and 84 are from the same case*)

Figure 83. Photomicrograph of a malignant diffuse type mesothelioma to demonstrate involvement of the stomach wall. Low power. A. F. I. P. Acc. No. 526647.

Figure 84. Higher magnification to demonstrate detail of involvement of stomach wall. Note tendency to form tubules. × 410. A. F. I. P. Acc. No. 526647.

*From the Armed Forces Institute of Pathology.

Fig. 83

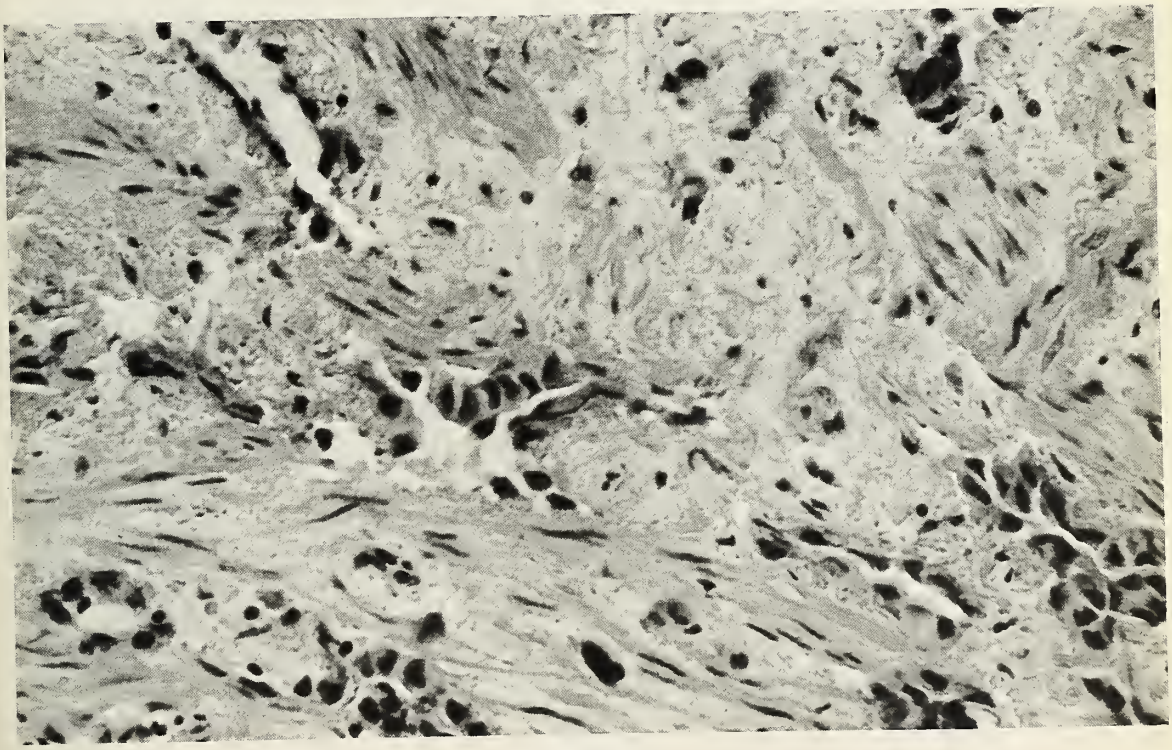

Fig. 84

MESOTHELIOMA

(Figures 85 and 86 are from the same case)

Figure 85. Localized malignant mesothelioma invading the ileum to the submucosa. × 130. A. F. I. P. Acc. No. 219023–18.

Figure 86. The tumor cells are arranged in solid masses and tend to form tubules. × 500. A. F. I. P. Acc. No. 219023–19.

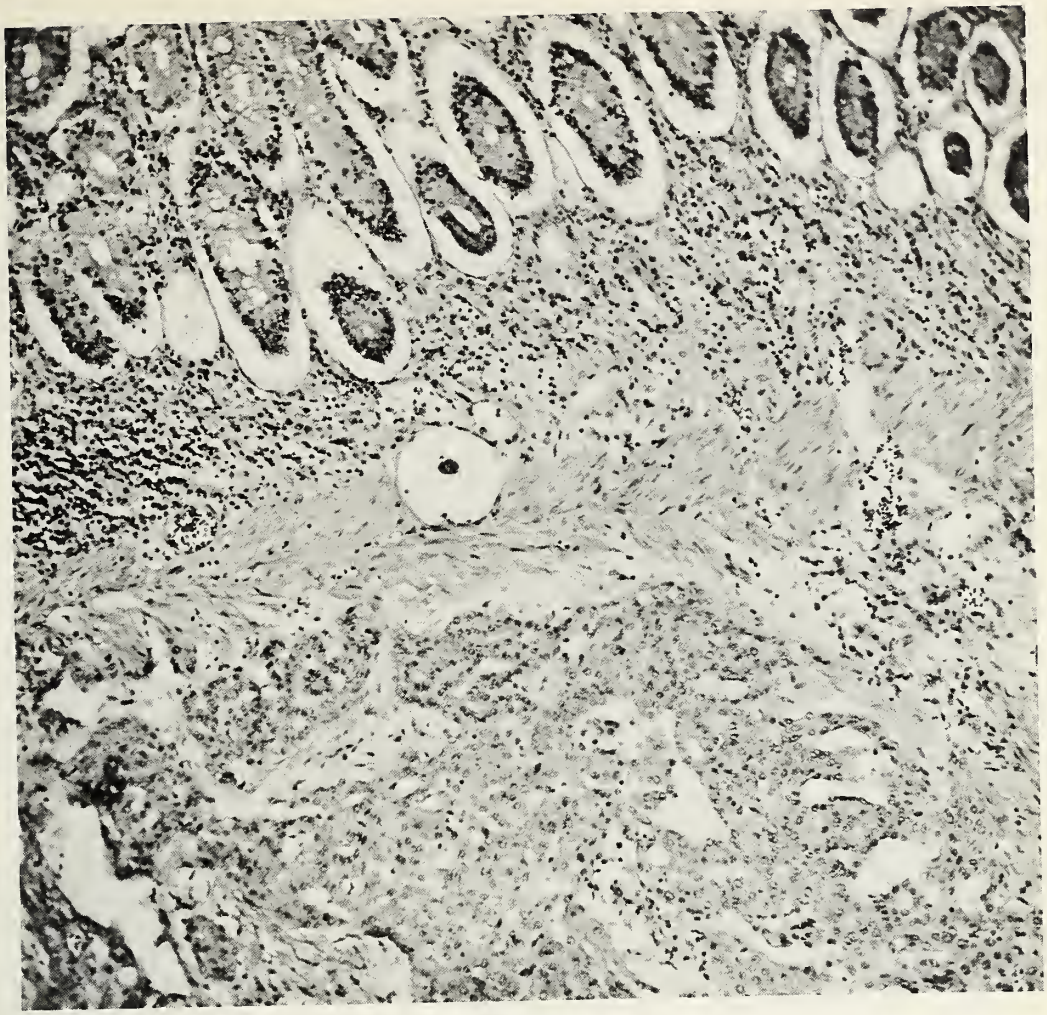

Fig. 85

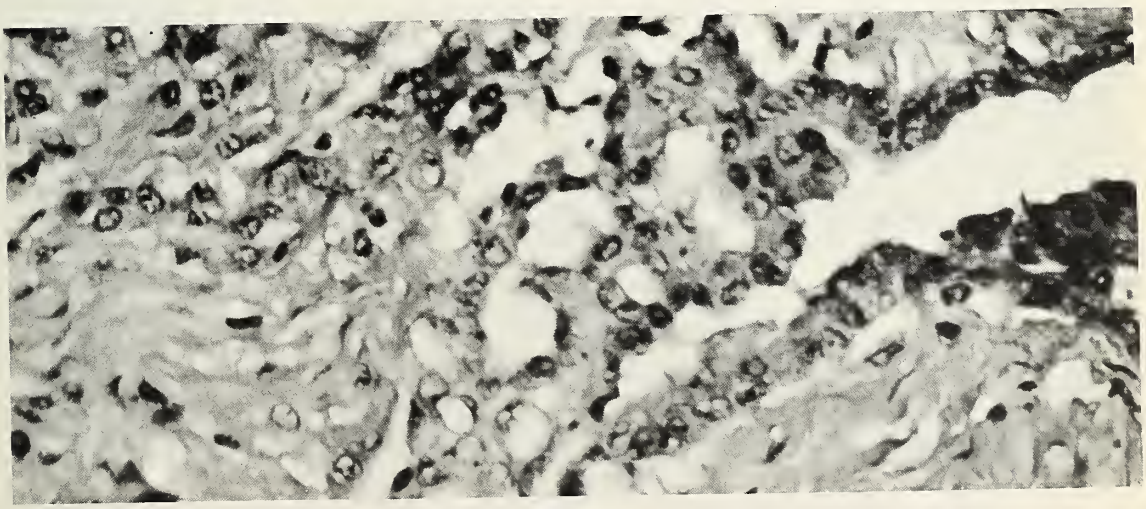

Fig. 86

MESOTHELIOMA

(Figures 87 and 88 are from the same case*)

Figure 87. Photomicrograph of localized malignant tubular mesothelioma. × 545. A. F. I. P. Acc. No. 219023–15.

Figure 88. Photomicrograph of malignant localized mesothelioma, primary sarcomatous-like area. × 545. A. F. I. P. Acc. No. 219023–16.

*Case courtesy of Dr. Roger Terry, Strong Memorial Hospital, New York, N. Y., and Dr. Frank McKee, Genesee Hospital, Rochester, N. Y.

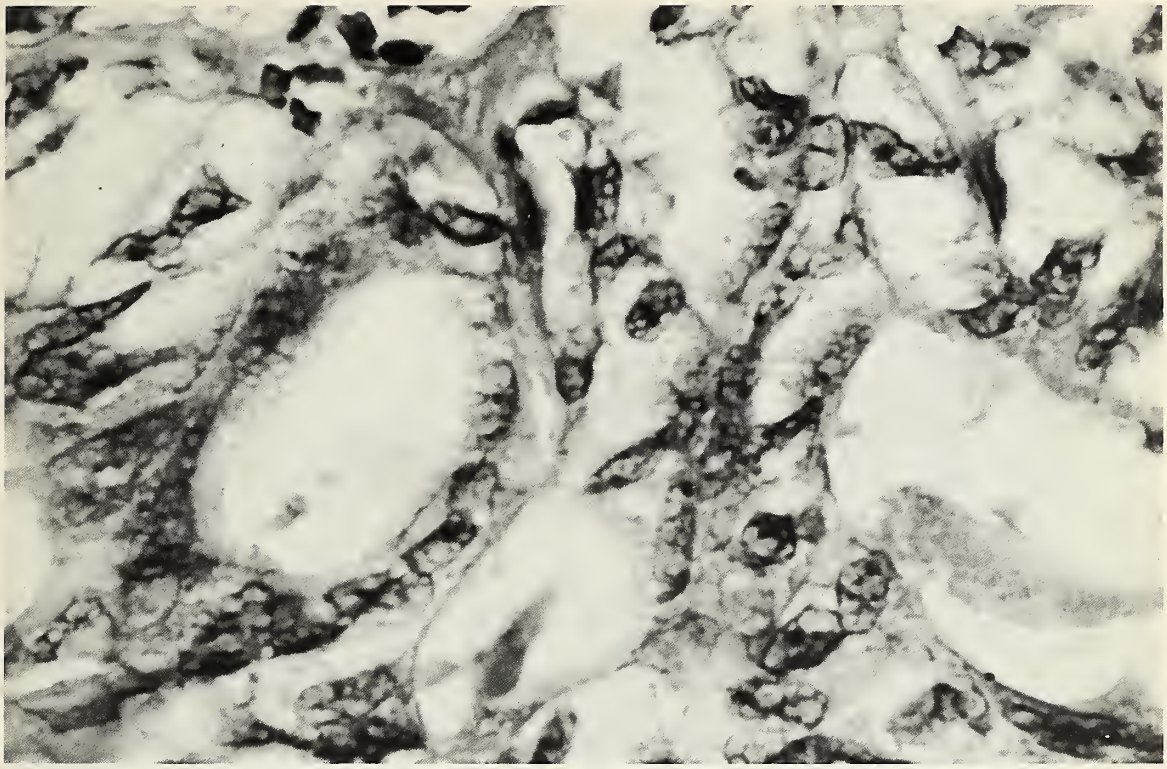

Fig. 87

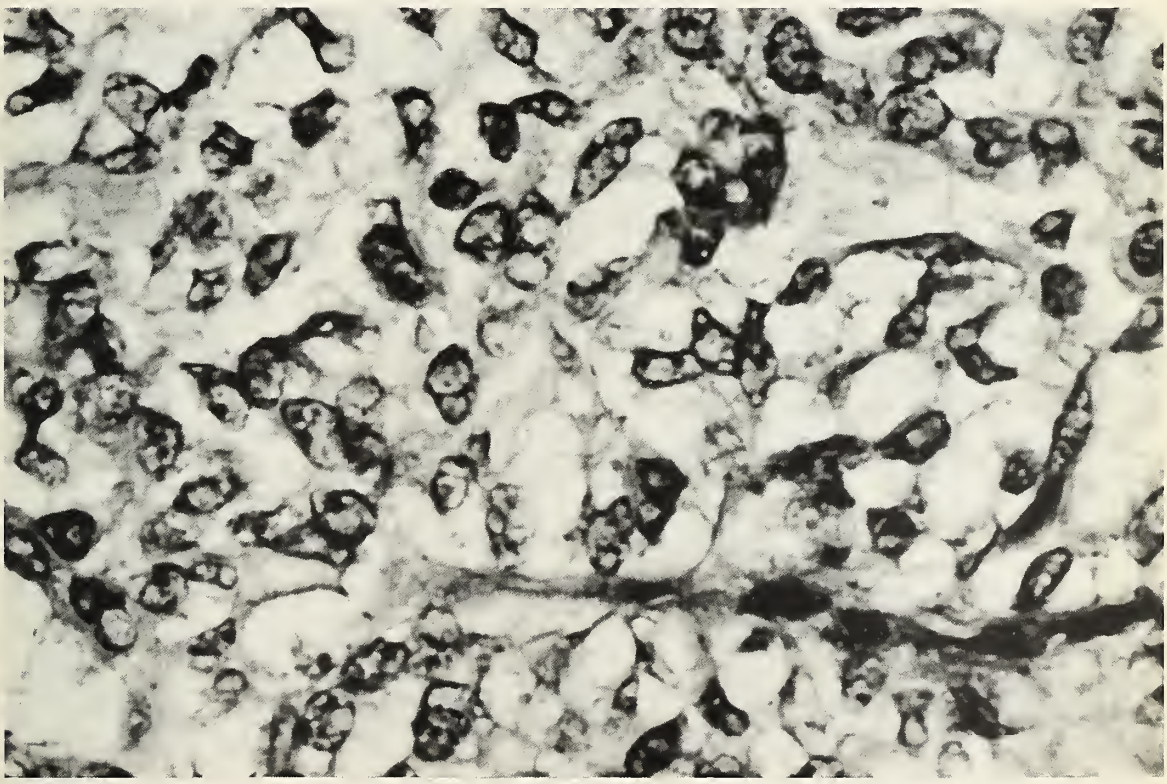

Fig. 88

METASTATIC CARCINOMA OF OVARY

(Figures 89 and 90 are from the same case*)

Figure 89. Photomicrograph of a peritoneal metastasis from a serous cystadenoma of the ovary. Note the similarity to primary mesothelioma. × 240. A. F. I. P. Acc. No. 314863–24013.

Figure 90. Photomicrograph from another area of the metastatic tumor. × 240. A. F. I. P. Acc. No. 314863–24012.

*Case of Dr. James R. Lisa, Doctor's Hospital, New York, N. Y. Used by Dr. A. P. Stout, figure 27, Case 19, Seminar on Tumors of the Soft Tissues, American Society of Clinical Pathologists, Chicago, Ill., 1951.

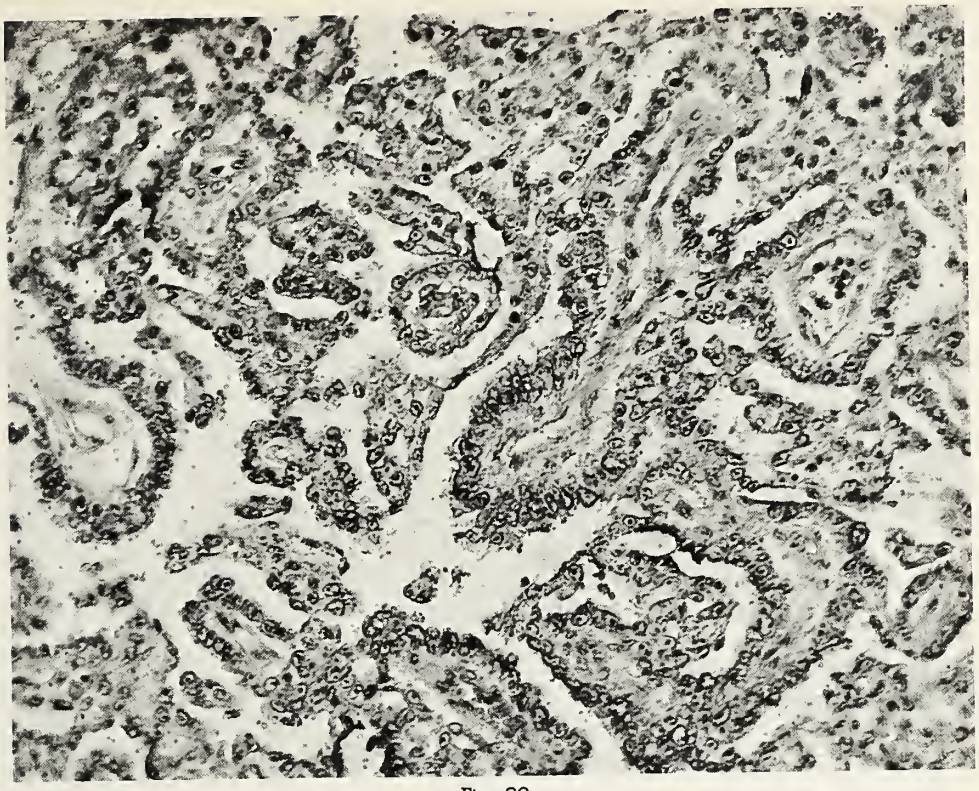

Fig. 89

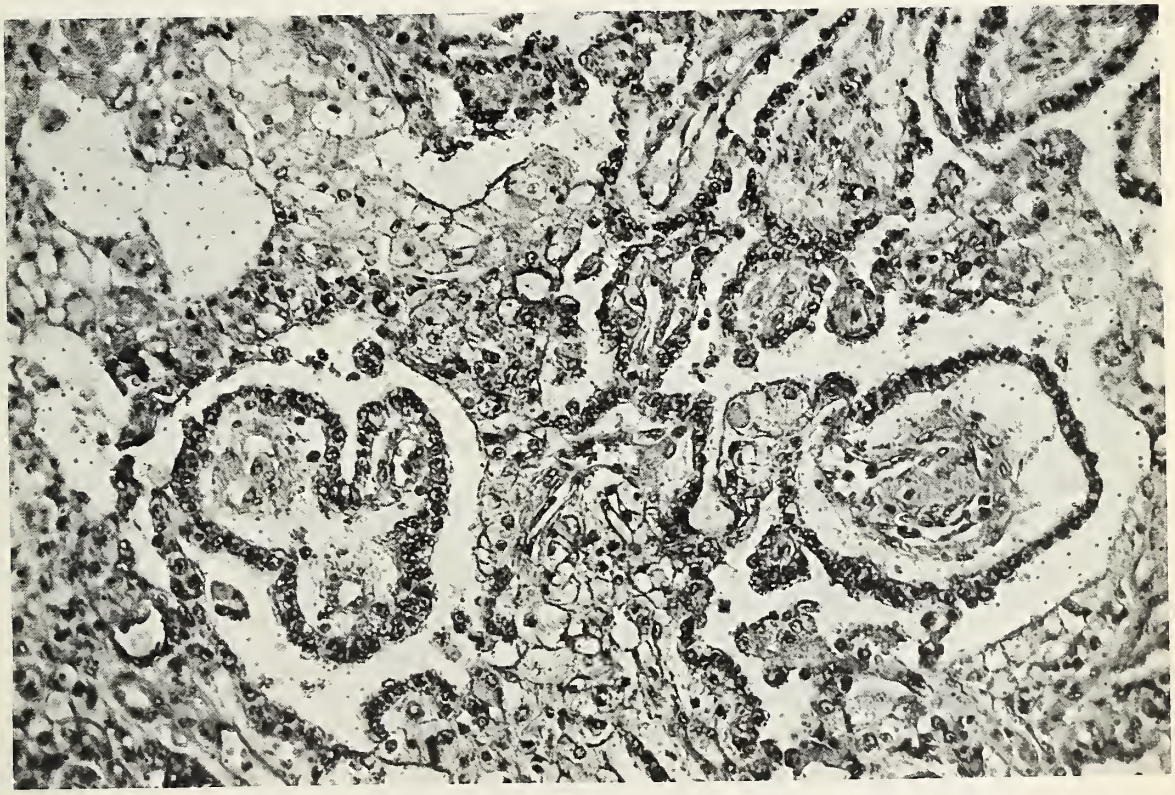

Fig. 90

METASTATIC CARCINOMA

(Figures 91 and 92 are from the case*)

Figure 91. Typical metastatic carcinoma involving the peritoneum. A. F. I. P. Acc. No. 219023–86.

Figure 92. Higher magnification of the metastatic nodules. A. F. I. P. Acc. No. 219023–87.

*Courtesy of Dr. Richard Johnson, Ellis Fischel State Cancer Hospital, Columbia, Mo.

Fig. 91

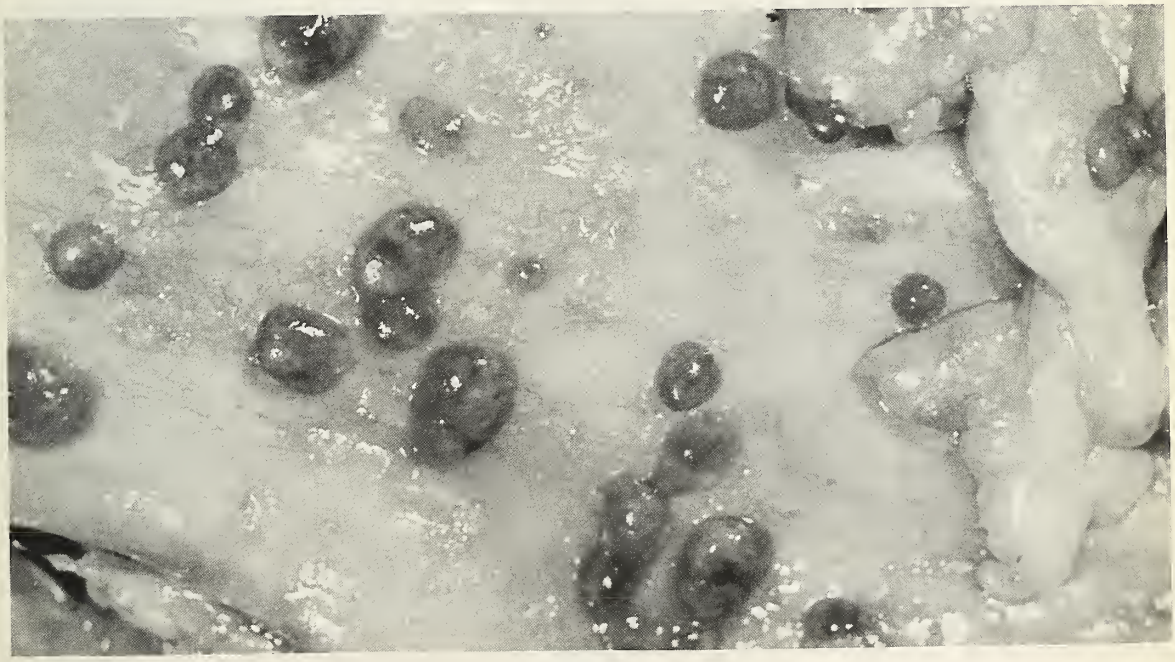

Fig. 92

METASTATIC CARCINOMA

Figure 93. Metastatic squamous carcinoma diffusely involving the peritoneum. A. F. I. P. Acc. No. 219023–88.

FOCAL PROLIFERATION PERITONEUM

Figure 94. Focal proliferation of the peritoneum showing the formation of pseudoacini. This was seen grossly as a small gray area of thickening of the peritoneum. × 240. A. F. I. P. Acc. No. 219023–104.

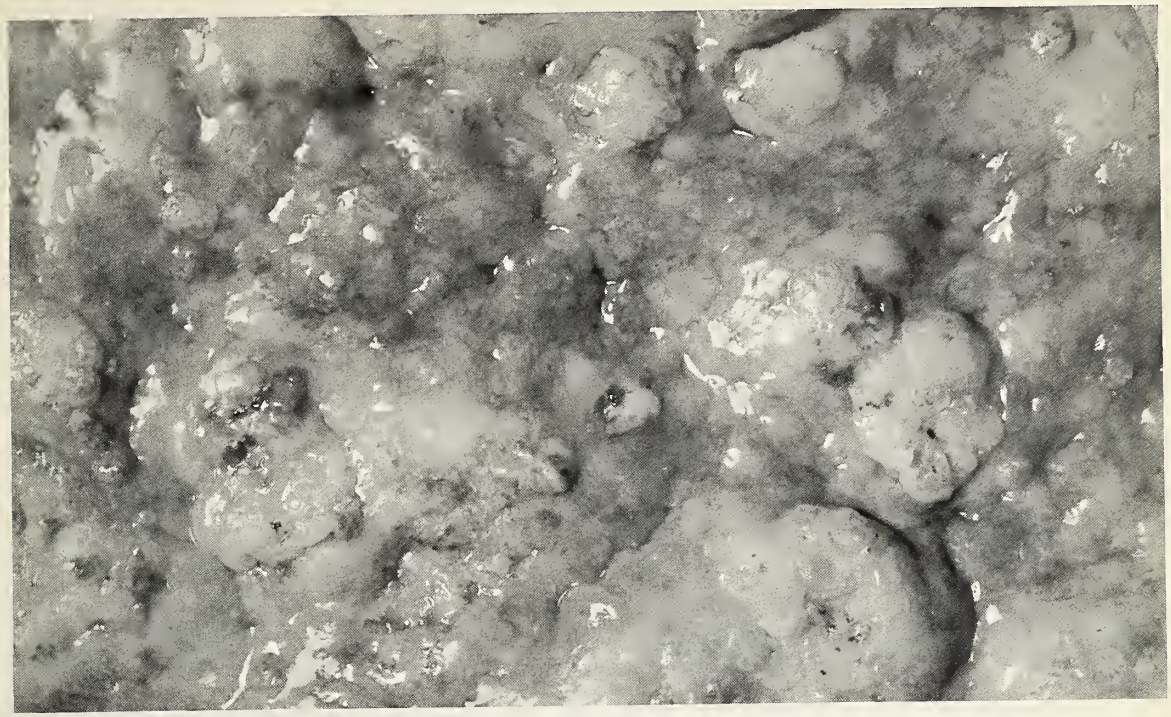

Fig. 93

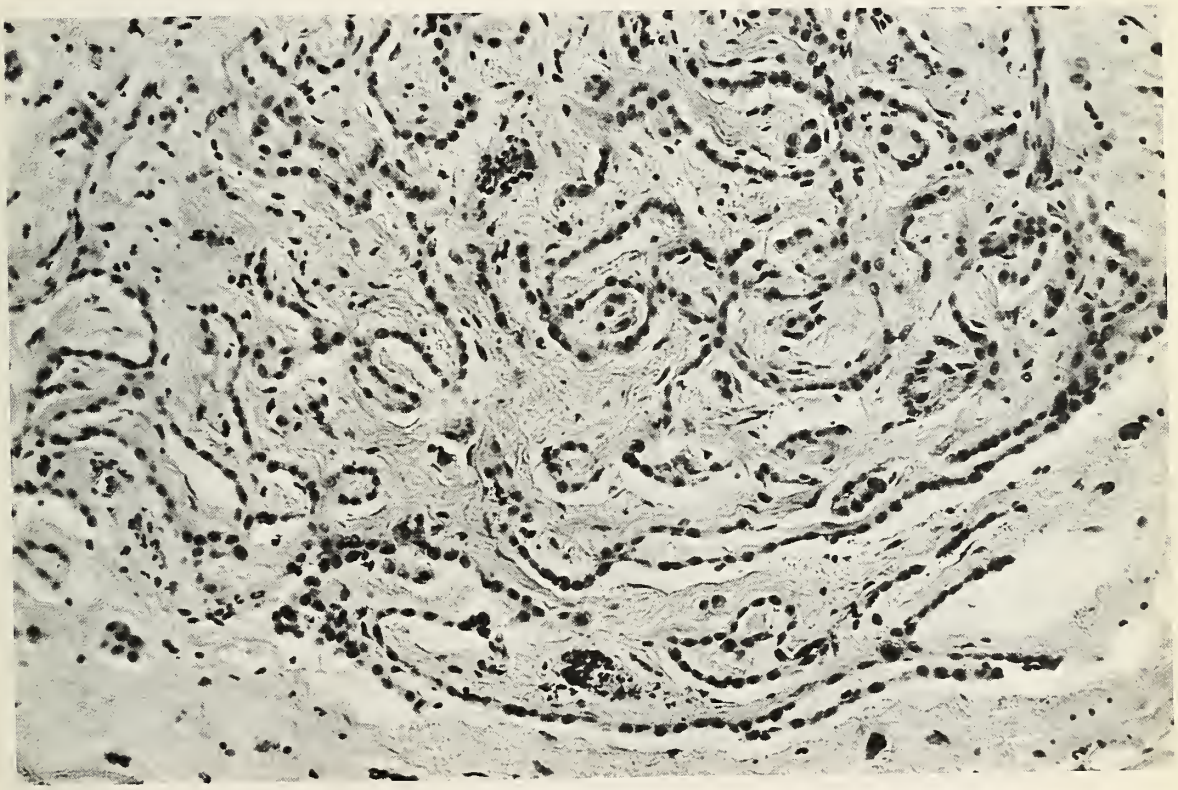

Fig. 94

ASCITIC FLUID

Figure 95. Large clump of mesothelial cells in ascitic fluid sediment, arranged in the form of pseudoacini, mistaken for metastatic carcinoma. Patient had cirrhosis of the liver, proven at autopsy. × 100. A. F. I. P. Acc. No. 219023–97.

Figure 96. Clumps of mesothelial cells with the form of pseudoacini. This patient did not have carcinoma. × 600. A. F. I. P. Acc. No. 219023–98.

Figure 97. A normal mesothelial cell in mitosis. × 1530. A. F. I. P. Acc. No. 219023–99.

Fig. 95

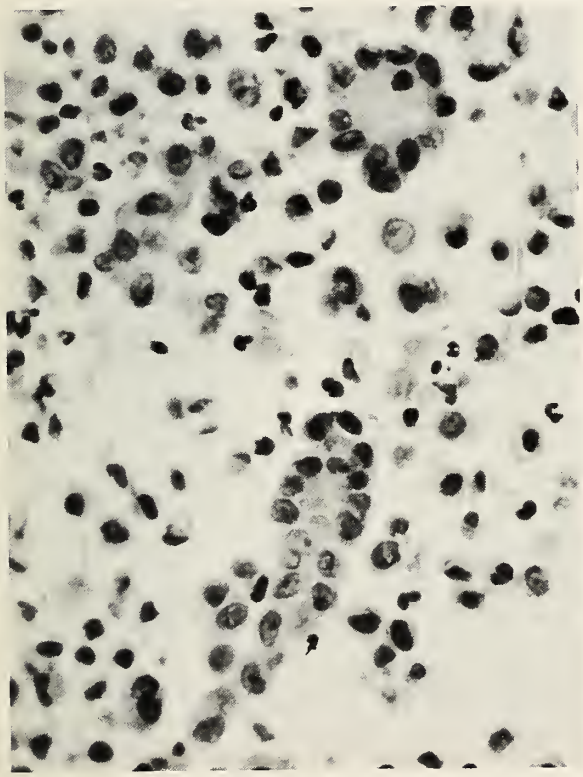

Fig. 96

Fig. 97

ASCITIC FLUID

Figure 98. Signet ring cells containing mucin are present. This patient had a primary neoplasm of the large bowel with metastases to the peritoneal surface. × 900. A. F. I. P. Acc. No. 219023–96.

Figure 99. Mesothelial cells in ascitic fluid. Note false signet ring type easily confused with carcinoma. The material within the cytoplasm is probably fat. × 900. A. F. I. P. Acc. No. 219023–95.

Figure 100. Single neoplastic cells with prominent deep-staining nuclei. Cytoplasm scanty. Note the large size of the nuclei as compared with those in the non-neoplastic mesothelial cells in figure 99. × 900. A. F. I. P. Acc. No. 219023–92.

Figure 101. Single neoplastic cells with prominent deep-staining nuclei in stained section of stomach. × 900. A. F. I. P. Acc. No. 219023–93.

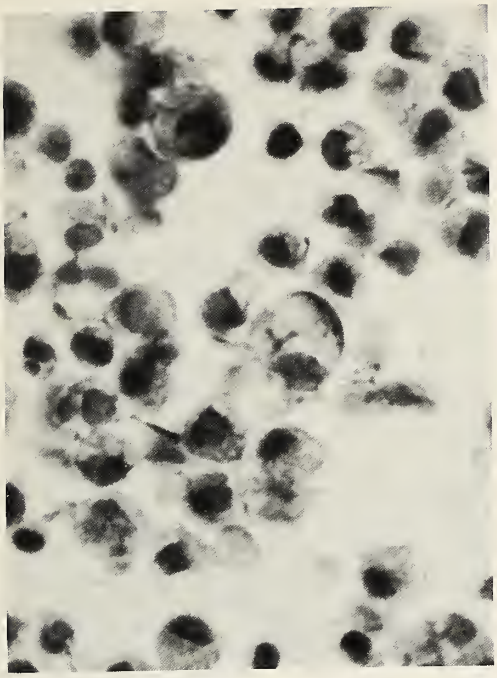

Fig. 98

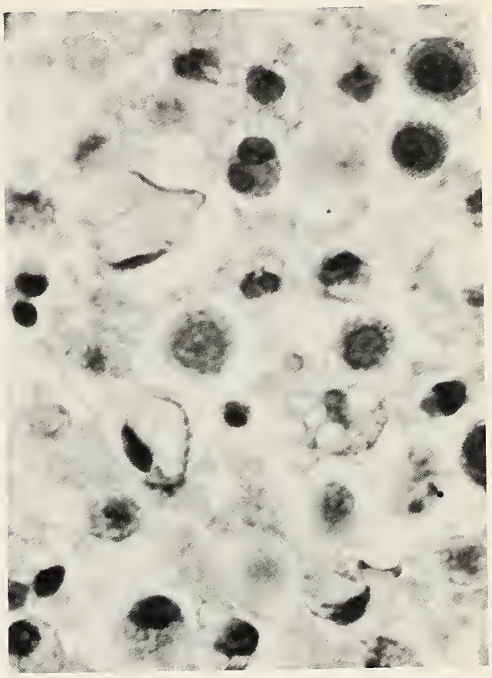

Fig. 99

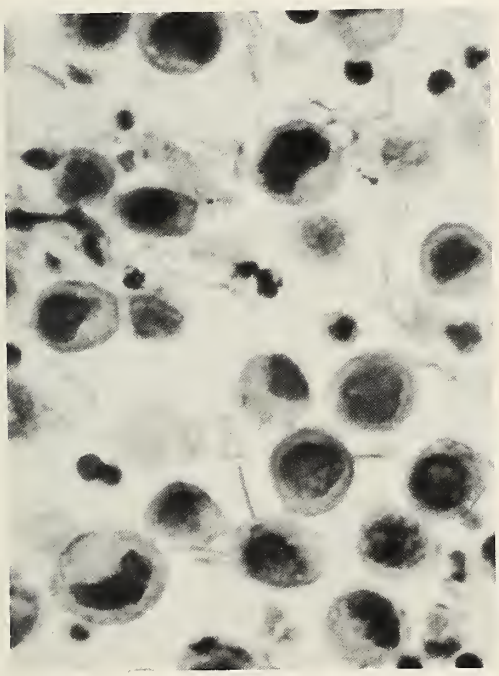

Fig. 100

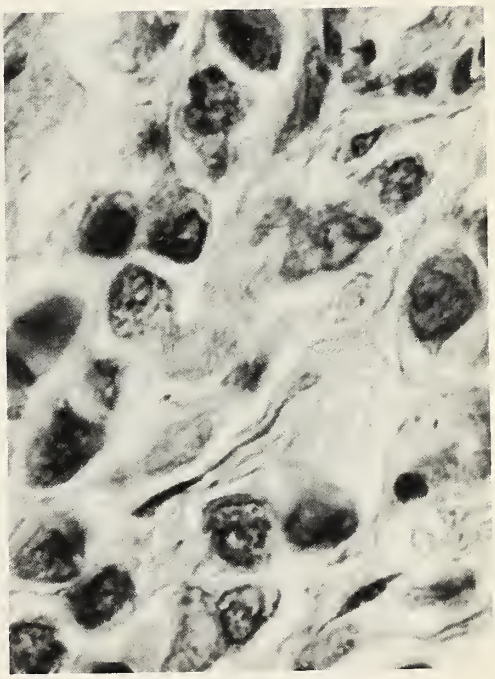

Fig. 101

ASCITIC FLUID

(Figures 102–104 are from the same case)

Figure 102. Serous adenocarcinoma of the ovary. Stained ascitic fluid sediment showing clusters of tumor cells. Note prominent nuclei and atypical mitotic figure. × 900. A. F. I. P. Acc. No. 219023–89.

Figure 103. Smear of centrifuged ascitic fluid sediment. The nuclei and nucleoli are prominent. × 900. A. F. I. P. Acc. No. 219023–90.

Figure 104. Section of ovarian tumor. Note resemblance of individual tumor cells to those seen in stained smear and sectioned stained sediment. × 900. A. F. I. P. Acc. No. 219023–91.

Figure 105. Clump of tumor cells arranged in an acinus in ascitic fluid sediment. Note prominent nuclei and atypical mitotic figure. × 900. A. F. I. P. Acc. No. 219023–94.

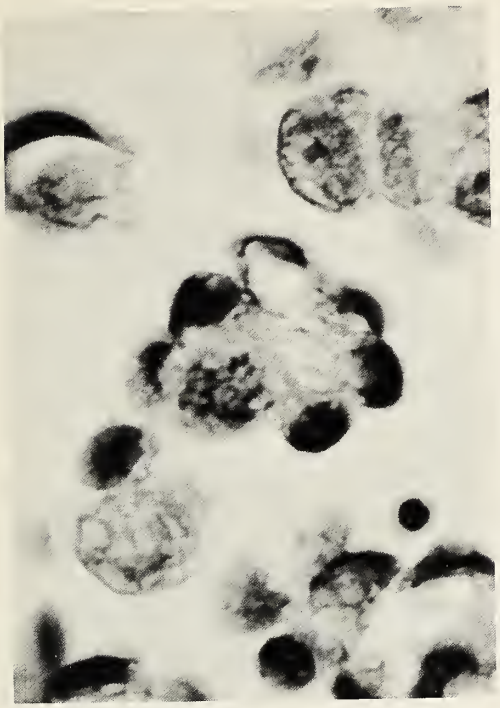

Fig. 102

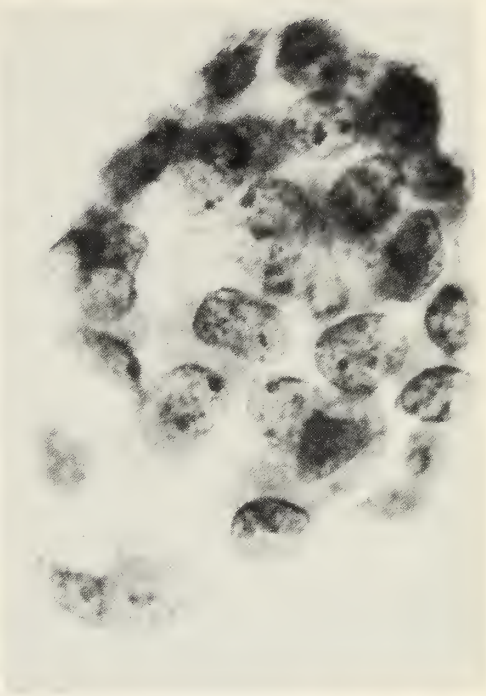

Fig. 103

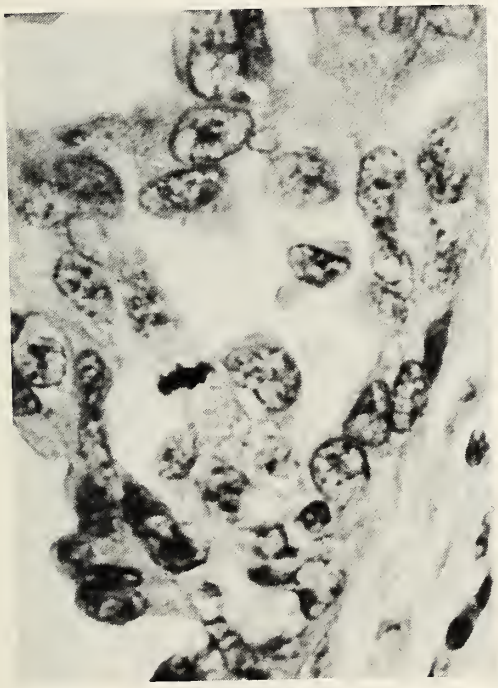

Fig. 104

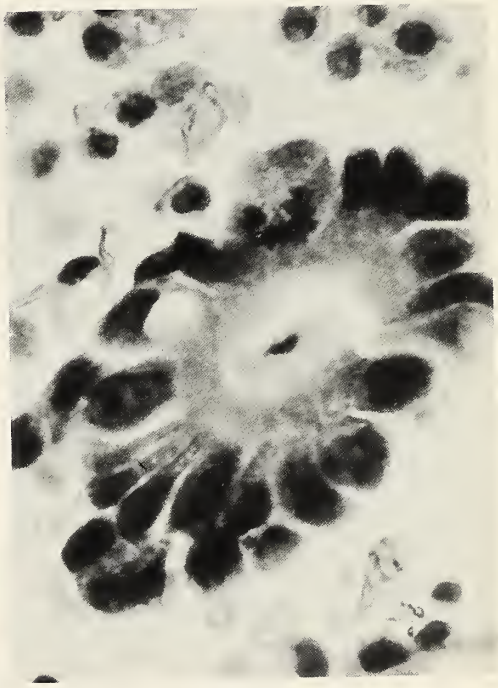

Fig. 105

Sano, Weiss, and Gault). Histochemical studies of production of hyaluronic acid may be confirmatory. Benign variants exist (Wells; Yoshida; Sternberg). It is the majority opinion that although primary peritoneal tumors are rare, they do exist.

Pseudomyxoma peritonei represents metastatic implants on the peritoneal surface from either a carcinoma of the appendix or a pseudomucinous carcinoma of the ovary. At times there may be changes in both organs. There has been considerable discussion concerning the pathogenesis of this condition. Experimental attempts to transplant the normal lining epithelium of the appendix to the peritoneum have been successful. However, if the relatively uninfected appendix is laid open, the secreted mucus acts as a foreign body and peritoneal nodules will form (Bergan). It appears logical that if a mucocele of the appendix forms, expands, and ruptures, the material will act in a similar fashion; but if the appendix is removed, the mucinous masses of foreign material will also disappear (Woodruff and McDonald). Metastatic masses on the peritoneal surface from mucus-secreting adenocarcinomas of the appendix or ovary have an autonomous growth. Removal of the ovary or appendix will not affect the growth of the metastases. These metastatic gelatinous masses have been likened to "frog's spawn" and in time may fill the entire abdomen ("jelly belly"). These masses kill by local infiltration of bowel, abdominal wall, and bladder, and may cause intestinal obstruction (Chaffee and LeGrand). Attempts to remove these masses often result in fatal peritonitis. Distant metastases are rare. Microscopically, large masses of mucus and well differentiated glands are present.

Peritoneal metastases can show various forms including discrete nodules, plaquelike masses, and cystic forms (figs. 91–93). The gross pattern is influenced by the amount of connective tissue, mucin production, and microscopic type. At times lymphosarcoma forms large cellular fish-flesh nodules on the peritoneal surface. The commonest sources of such metastatic lesions are the ovary, stomach, large bowel, and body of pancreas. Primary carcinomas from almost any organ can involve the peritoneum. Metastatic carcinoma of the peritoneum can be simulated by tuberculosis (Daniel), talcum powder granuloma (Seelig; Seelig and Verda), rupture of a dermoid cyst of the ovary (Auer, Dockerty, and Mayo), encapsulated food particles from a perforated gastric ulcer (Daniel).

Recognition of Tumor Cells in Ascitic Fluid

The ability to recognize tumor cells in ascitic fluid may obviate an exploratory laparotomy. We study the fluids received in our laboratory by two techniques. In one the fluid is centrifuged and the sediment blocked and sectioned at various levels, and in the other smears are made and stained by the Papanicolaou technique. This latter method better preserves cytologic detail. The two methods complement each other, but we have found the smear method extremely valuable. Saphir has been able to reach a high degree of

accuracy in examining both sediment and smears. Such a high degree of accuracy requires considerable background and experience.

Errors in this method of examination are legion. Frequent errors are made on the side of the false positive diagnosis. These errors are related to the consummate ability of mesothelial cells to simulate carcinoma. For instance, in the presence of inflammation and cirrhosis of the liver extreme proliferation of mesothelial cells may occur (fig. 94). Apparent acini may occur when a sheet of mesothelial cells doubles on itself (Saphir; figs. 95, 96). Mesothelial cells may have multiple nuclei or even mitotic figures (fig. 97). The cytoplasm is pale pink and the nucleoli are poorly defined. Furthermore, signet ring type cells may lead one to suspect metastatic carcinoma (fig. 98). These cells, however, do not necessarily take the stain for epithelial mucin (fig. 99). We have made an unequivocal diagnosis of malignant tumors under certain circumstances. In a few instances large clumps of obviously malignant tumor cells are present (figs. 102, 103, 105). At other times the diagnosis can be made more on the findings of multiple single tumor cells (fig. 100). This diagnosis is usually made on the basis of the nuclear changes. The nuclei are large, deep-staining, and often have prominent nucleoli. Mitotic figures are significant only if they are atypical. The nucleus in a malignant cell is often twice the size of the nucleus of the mesothelial cells. Prominent nucleoli are also significant.

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